<table>
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<th>Title</th>
<th>Dentofacial growth in patients with Sotos syndrome</th>
</tr>
</thead>
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<tr>
<td>Author(s)</td>
<td>Takei, K; Sueishi, K; Yamaguchi, H; Ohtawa, Y</td>
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</table>
Case Report

Dentofacial Growth in Patients with Sotos Syndrome

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Abstract

Sotos syndrome is an overgrowth syndrome leading to peculiar facial characteristics, large hands and feet, and mental retardation. The maxillofacial characteristics are metopic protrusion, a high and narrow palate and a tapered mandible. In this study, we evaluated changes in maxillofacial growth in 2 patients with cerebral gigantism during the peri-pubertal period.

Patient 1 was a boy aged 8 years at the first examination. The face showed midface retraction and a tapered mandible.

Maxillary median diastema with an OJ of 2.5 mm and OB of 1.0 mm was observed, and the molar region showed mandibular mesial occlusion. Radiography revealed a lack of 15, 25, 37, 47, 14, 24, 34 and 44. Cephalometrics demonstrated maxillary and mandibular retrusion with an SNA of 68° and an SNB of 70°, and the patient had leptoprosopia with a mandibular plane of 38.0°. This plane was 45° at the time of re-examination when the patient was 14 years old, showing an increase in the lower facial height and decreases in facial axis and depth. Patient 2 was a boy aged 14 years at the first examination. The face showed mandibular retrusion and tapering. The occlusion was angle class II div. 1, OJ 14 mm, and OB −1 mm. Cephalometrics demonstrated maxillary and mandibular retrusion with an SNA of 74.5° and an SNB of 69.5°, and the patient had leptoprosopia with a mandibular plane of 37.0°. At the time of re-examination, when the patient was 16 years old, the mandibular plane was 42.5°, showing an increase in lower facial height and decreases in facial axis and depth.

In this syndrome, excessive facial height without mandibular forward overgrowth is observed. Since the facial height tended to increase by growth during the peripubertal period, maxillofacial vertical growth is considered important in the treatment of this syndrome.

Key words: Sotos syndrome—Cephalometrical radiographs—Vertical growth pattern

Objectives

Cerebral gigantism (Sotos syndrome), showing heavy body weight at birth and excessive growth over 4 years after birth, is an overgrowth syndrome characterized by bone age acceleration, specific facial characteristics such as mandibular prognathism, large hands
and feet, valgus and mental retardation, and was first reported in 1964 by Sotos et al.\textsuperscript{7).} The maxillofacial characteristics are megacephaly, dolichocephalia, metopic protrusion, anterior fontanel dilation, strabismus, ocular hypertelorism, a high and narrow palate, a tapered mandible, and early eruption of teeth\textsuperscript{6).} There have been studies on maxillofacial growth and pronunciation in infants with this syndrome\textsuperscript{5),} but no studies during the peripubertal period were found. In this study, we evaluated changes in maxillofacial growth in 2 patients with cerebral gigantism during the peripubertal period.

![Diagram of maxillofacial growth analysis](image)

**Fig. 1** Morphological analysis of maxillofacial skull (Susami et al.\textsuperscript{8})

A) Calvarial region: H, maximal cranial height; L, maximal cranial major diameter; Ha, anterior cranial height in N region; Hp, posterior cranial height in Bz region; W, maximal cranial width; Cranial indices, H/L, H/W, W/L, Hp/Ha; Cranial capacity index, H + L + W/3.

B) Basicranial region: S-N, cranial major diameter; S-Ba, major diameter of posterior skull; S-ER', sphenoidal major diameter in anterior skull base; S-SOS', sphenoidal major diameter in posterior skull base; NSBa, basicranial angle; S-ER'/S-N, sphenoidal ratio in anterior skull base; S-SOS'/S-Ba, sphenoidal ratio posterior skull base; S-Ba/S-N, anteroposterior ratio in skull base.

C) Maxillary region: N-ANS, anterior maxillary height; S-PNS, posterior maxillary height; ANS-PNS, anteroposterior major diameter in upper jaw; S-PNS/N-ANS, anteroposterior ratio of maxillary height.

D) SN-IOR, protrusion of orbital anterior margin; BIOD, interorbital bone distance; BIOD/W, ratio of interorbital bone distant to cranial width.

E) Ar-Go, mandibular ramus height; Go-Pog, mandibular corpus length; Ar-Pog, mandibular length; Ar-Go/Go-Pog; ratio of mandibular ramus height to mandibular corpus length.

A) Shows diagram of how evaluation was performed for length of skull, width, diameter, and ratio according to radiography of profile and frontal image.

B) Shows diagram of how evaluation was performed for length of base of skull, and ratio according to radiography of profile.

C) Shows diagram of how evaluation was performed for maxillary positional relationship according to radiography of profile.

D) Shows diagram of how evaluation was performed for length of eye socket, and positional relationship according to radiography of profile and frontal image.

E) Shows diagram of how evaluation was performed for length of maxillary bone, and positional relationship according to radiography of profile.
Case Report

This study was performed using frontal and lateral cephalometrical radiographs taken at the first examination and re-examination in 2 patients with Sotos syndrome who consulted the Orthodontic Department of Suidobashi Hospital, Tokyo Dental College. Ricketts analysis, Downs & Northwestern analysis, and analysis of maxillofacial skull morphology by Susami et al.8) (Fig. 1) were used to evaluate the radiographs.

Patient 1 was a boy aged 8 years 6 months at the first examination. His height was 142.2 cm, body weight 37 kg, and bone age 11.0 years. The patient consulted our hospital with chiefly for median diastema and a lack of teeth. The face was symmetric, with midface retraction and a tapered mandible (Fig. 2). In the oral cavity, maxillary median diastema with an OJ of 2.5 mm and OB of 1.0 mm was observed, and the molar region was angle II. Eruption of the first maxillary molar was delayed (Fig. 2). Radiography revealed a lack of bilateral second premolars and molars in the lower jaw and bilateral first premolars in the upper and lower jaws.

Ricketts analysis of the profile obtained by radiographic cephalometry (Tables 1-1, 1-2, and 1-3) demonstrated that convexity (−4.5°) was small, showing mandibular prognathism, and the facial axis (82.0°), facial depth (83.0°), mandibular plane (37.5°), and lower facial height (51.0°) were large, revealing a dolichofacial pattern.

Downs & Northwestern analysis indicated that the SNA (68°) and SNB (70°) were small, showing maxillary and mandibular retrusion. It was also noted that the mandibular plane (38.0°) and gonial angle (133.5°) were large, and that the ramus angle (80°) was small, showing a high angle.

Analysis by the method of Susami et al.8) demonstrated that, in the calvarial region, maximal cranial height (H, 160 mm), maximal cranial major diameter (L, 216 mm), posterior cranial height in the Ba region (Hp, 158.5 mm), and cranial capacity index (H(L+W)/3, 181) were large, revealing megacephaly and leptoprosopia. In the basi-cranial region, the major diameter (S-N, 76 mm) was long, showing leptoprosopia. Anterior maxillary height (N-ANS, 61 mm) was large in the maxillary profile, and interorbital bone distance (BIOD, 26.5 mm) was also large on the frontal maxillary image. In the mandibular profile, the mandibular corpus length (Go-Pog, 75.5 mm) and mandibular length (Ar-Pog, 109 mm) were large, showing a high angle.

At the time of re-examination (Figs. 3 and 4, Tables 1-1, 1-2, and 1-3), the patient was 14 years 6 months old.

Ricketts analysis demonstrated that the facial axis and depth had decreased to 80° and 82°, respectively, and that the mandibular plane and lower facial height had markedly increased to 40° and 53°, respectively, revealing an increase in the dolichofacial pattern.

Downs & Northwestern analysis indicated that the gonial angle had increased to 136°, showing an increase in the high angle.
<table>
<thead>
<tr>
<th>Clinical norm</th>
<th>8y6m</th>
<th>Clinical norm</th>
<th>14y6m</th>
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<tr>
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<td>82</td>
<td>86</td>
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<tr>
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<td>37.5</td>
<td>29</td>
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<td>Lower facial height</td>
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<td>51</td>
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<td>Intercanine width</td>
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<tr>
<td>Facial width</td>
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Ricketts analysis demonstrated that the facial axis and depth had decreased to 80° and 82°, respectively, and that mandibular plane and lower facial height markedly increased to 40° and 53°, respectively, revealing increase in dolichofacial pattern.

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<th>Clinical norm</th>
<th>14y6m</th>
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<tr>
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<td>F-H to SN plane</td>
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Downs & Northwestern analysis indicated that gonial angle had increased to 136°, showing increase in high angle.
Table 1-3  Susami et al. analysis

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<tr>
<td>Profile a standard on N-Ba plane</td>
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<tr>
<td>H; Maximal cranial height</td>
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<td>160</td>
<td>156.1</td>
<td>161</td>
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<td>L; Maximal cranial major diameter</td>
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<td>158.5</td>
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<td>Frontal image</td>
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<tr>
<td>W; Maximal cranial width</td>
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<td>168.3</td>
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<td>H + L + W/3</td>
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<td>Profile</td>
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<td>S-N; Cranial major diameter</td>
<td>64.8</td>
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<tr>
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<td>43</td>
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<td>N-ANS; Anterior maxillary height</td>
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<td>S-PNS; Posterior maxillary height</td>
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<tr>
<td>SN-IOR; Protrusion of the orbital anterior margin</td>
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<tr>
<td>Frontal image</td>
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<tr>
<td>BIOD; Interorbital bone distance</td>
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<td>23.7</td>
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<tr>
<td>BIOD/W; Ratio of the interorbital bone distance to the cranial width</td>
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<td>0.159</td>
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<td>E) Mandibular region</td>
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<td>Profile</td>
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<tr>
<td>Ar-Go; Mandibular ramus height</td>
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<tr>
<td>Go-Pog; Mandibular corpus length</td>
<td>66.4</td>
<td>75.5</td>
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<tr>
<td>Ar-Pog; Mandibular length</td>
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<td>123</td>
<td>117</td>
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<tr>
<td>Gonial angle</td>
<td>128.1</td>
<td>139</td>
<td>123.4</td>
<td>138</td>
</tr>
<tr>
<td>Mandibular plane-SN, SNB</td>
<td>76.1</td>
<td>70</td>
<td>79</td>
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</tr>
<tr>
<td>Ar-Go/Go-Pog; Ratio of the mandibular ramus height to the mandibular corpus length</td>
<td>0.65</td>
<td>0.57</td>
<td>0.73</td>
<td>0.613</td>
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</table>

Analysis by method of Susami et al. demonstrated that maximal cranial major diameter, mandibular ramus height, and cranial capacity index had increased to 220.5mm, 49mm (43mm at first examination), and 183.5, respectively.
Analysis by the method of Susami et al. demonstrated that the maximal cranial major diameter, mandibular ramus height, and cranial capacity index had increased to 220.5 mm, 49 mm (43 mm at the first examination), and 183.5, respectively.

Figure 4 shows the profile produced by superimposing the profile obtained at the first examination on that obtained at re-examination at the S point on the S-N plane, and the frontal image produced by superimposing the image obtained at the first examination on that obtained at re-examination at Cock’s comb on the maxillofacial median. As in this figure, the skull and mandibular bone grew vertically, and a high angle was observed.

Ricketts 5-position analysis (Fig. 5) demonstrated a 2° dilation of the facial axis by the first superimposition, a 1.5-mm retraction of point A by the second superimposition, a 1-mm inclination of the maxillary medial incisor to the lip and a 1-mm mesial movement of the first maxillary molar by the third superimposition, a 2-mm inclination of the mandibular medial incisor to the tongue and a 1-mm mesial movement of the first mandibular molar by the fourth superimposition, and a
1.5-mm retraction of the lower lip with respect to the E-line by the fifth superimposition.

Patient 2 was a boy aged 14 years 6 months at the first examination. His height was 174 cm, body weight 50.0 kg, and bone age 14.8 years. Mental retardation was observed. The patient consulted our hospital chiefly for maxillary anterior tooth protrusion. The face was symmetric, and showed mandibular retrusion and tapering (Fig. 6). The occlusion was angle class II div. 1, OJ 14 mm, and OB −1 mm (Fig. 6).

Ricketts 10-factor analysis of the profile obtained by radiographic cephalometrics (Tables 2-1, 2-2, and 2-3) demonstrated that the facial axis (87°), facial depth (83.0°), mandibular plane (36°), lower facial height (57°), and cranial length (63.5°) were large, showing a high angle. Ricketts analysis of the frontal image demonstrated a small facial width (132°).

Downs & Northwestern analysis indicated that the mandibular plane (37°) and gonial angle (133°) were large, and that the ramus angle (84°), SNA (74.5°), and SNB (69.5°) were small, showing maxillary and mandibular retrusion.

Analysis by the method of Susami et al. demonstrated leptoprosopia, with a maximal cranial height (H) of 151 mm, maximal cranial major diameter (L) of 167 mm, anterior cranial height in the N region (Ha) of

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**Treatment Results**

**5 POSITION ANALYSIS**

<table>
<thead>
<tr>
<th>Name</th>
<th>Pre.</th>
<th>Post.</th>
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<tbody>
<tr>
<td>WM 2</td>
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<td></td>
<td>Red</td>
<td>18y1m</td>
</tr>
</tbody>
</table>

**SUMMARY OF TREATMENT RESULTS**

- S1: Fac. 2.5° open
- S2: Point A: 1.5° reduction
- S3: U1: 1mm flare
- S4: L1: no change
- S5: L2: 1mm loss
- S6: Lower Lip: 4mm from E-line base

**Fig. 5** Patient 1: 5-positions analysis

Ricketts 5-position analysis demonstrated 2° dilation of facial axis by first superimposition, 1.5-mm retraction of point A by second superimposition, 1-mm inclination of maxillary medial incisor to lip and 1-mm mesial movement of first maxillary molar by third superimposition, 2-mm inclination of mandibular medial incisor to tongue and 1-mm mesial movement of first mandibular molar by fourth superimposition, and 1.5-mm retraction of lower lip with respect to E-line by fifth superimposition.
109.5 mm, and posterior cranial height in the Ba region (Hp) of 143 mm in the calvarial profile, a maximal cranial width (W) of 164.5 mm on the frontal image, and a cranial major diameter (S-N) in the basicranial profile of 70 mm. In the maxillary profile, the anterior maxillary height (N-ANS) was 61.5 mm, posterior maxillary height (S-PNS) 51 mm, and anteroposterior maxillary diameter (ANS-PNS) 48.5 mm, and on the frontal image, the interorbital bone distance (BIOD) was 29.5 mm, showing ocular hypertelorism.

At the time of re-examination, the patient was 16 years 11 months old (Figs. 7 and 8, Tables 2-1, 2-2, and 2-3).

Ricketts 10-factor analysis demonstrated that the facial axis had decreased to 84°, showing a dolichofacial pattern. Ricketts analysis of the frontal image indicated that the maxillary width (63 mm), mandibular width (99 mm), and facial width (144 mm) were large.

Downs & Northwestern analysis revealed that the Y-axis was large (72°), revealing an increase in the high angle.

Analysis by the method of Susami et al. demonstrated that the maximal cranial major diameter was 185 mm in the calvarial profile, and the sphenoidal major diameter in the anterior skull base (S-ER') was 28.5 mm in the basicranial profile, indicating enhancement of the dolichofacial pattern. In the maxillary profile, the anterior maxillary height (N-ANS), posterior maxillary height (S-PNS), and anteroposterior major diameter in the upper jaw (ANS-PNS) had markedly increased to 63.5 mm, 52.5 mm, and 51 mm, respectively, and in the mandibular profile, the mandibular ramus height (Ar-Go) and mandibular length (Ar-Pog) had also increased to 48.5 mm and 113 mm, respectively, indicating an increase in the high angle.

Figure 8 shows the profile produced by superimposing the profile obtained at the first examination on that obtained at re-examination at the S point on the S-N plane, and the frontal image produced by superimposing the image obtained at the first examination on that obtained at re-examination at Cock’s comb on the maxillofacial median. As seen in this figure, the skull and mandibular bone had grown vertically grew, and a high angle was observed.

Ricketts 5-position analysis (Fig. 9) demonstrated a 2.5° dilation of the facial axis by the first superimposition, a 1.5-mm retraction of point A by the second superimposition, a 1-mm inclination of the maxillary medial incisor to the lip without any movement of the maxillary molars by the third superimposition, a 1-mm mesial movement of the mandibular molars without any movement of the mandibular anterior teeth by the fourth superimposition, and a 1-mm retraction of the lower lip with respect to the E-line by the fifth superimposition.

**Discussion**

In Sotos syndrome, as in other overgrowth syndromes such as Wiedeman-Beckwith syn-
Table 2-1  Ricketts 10-factor analysis

<table>
<thead>
<tr>
<th></th>
<th>Clinical norm</th>
<th>14y6m</th>
<th>Clinical norm</th>
<th>16y11m</th>
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</thead>
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<tr>
<td>Facial axis</td>
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<td>87</td>
<td>86</td>
<td>84</td>
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<tr>
<td>Facial depth</td>
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<td>81</td>
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<td>Mandibular plane</td>
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<td>28.8</td>
<td>41</td>
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<tr>
<td>Lower facial height</td>
<td>49</td>
<td>57</td>
<td>49</td>
<td>61</td>
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<tr>
<td>Mandibular arc</td>
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<tr>
<td>Convexity</td>
<td>2</td>
<td>3</td>
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<td>L1 to APO (distance)</td>
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<td>-3</td>
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<td>L1 to APO (degree)</td>
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<td>Lower lip E-plane</td>
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<td>Cranial deflection</td>
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<td>Porion location</td>
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<td>Corpus length</td>
<td>70.5</td>
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<td>73.5</td>
<td>68.7</td>
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<table>
<thead>
<tr>
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<th>Clinical norm</th>
<th>14y6m</th>
<th>Clinical norm</th>
<th>16y11m</th>
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<tbody>
<tr>
<td>Molar relation right</td>
<td>1.5</td>
<td>-1</td>
<td>1.5</td>
<td>-1</td>
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<tr>
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<td>Intermaxillary angle</td>
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<td>Max-Mand width right</td>
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<td>Molar to jaw right (Mand)</td>
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<td>Max. width</td>
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<tr>
<td>Mand. width</td>
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<td>Facial width</td>
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</table>

Ricketts 10-factor analysis demonstrated that facial axis had decreased to 84°, showing dolichofacial pattern. Ricketts analysis of frontal image indicated that maxillary width (63 mm), mandibular width (99 mm), and facial width (144 mm) were large.

Table 2-2  Downs & Northwestern analysis

<table>
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<th>Clinical norm</th>
<th>14y6m</th>
<th>16y11m</th>
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</thead>
<tbody>
<tr>
<td>Facial angle</td>
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<td>Convexity</td>
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<td>6</td>
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<td>Mandibular plane</td>
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<tr>
<td>Y axis</td>
<td>64</td>
<td>67.5</td>
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<tr>
<td>Occlusal plane</td>
<td>8.4</td>
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<tr>
<td>Interincisal</td>
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<tr>
<td>L-1 to occlusal</td>
<td>21.3</td>
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<tr>
<td>L-1 to mandibular</td>
<td>97.1</td>
<td>76</td>
<td>80</td>
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<tr>
<td>U-1 to A-P plane</td>
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<td>F-H to SN plane</td>
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<td>SNA</td>
<td>83.4</td>
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<td>SNB</td>
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<td>U-1 to FH plane</td>
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<tr>
<td>L-1 to FH plane</td>
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<tr>
<td>Gonial angle</td>
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<tr>
<td>Ramus angle</td>
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<td>85</td>
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</table>

Downs & Northwestern analysis revealed that Y axis was large (72°), revealing increase in high angle.
Table 2-3 Susami et al. analysis

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<tbody>
<tr>
<td>A) Calvarial region</td>
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<tr>
<td>Profile a standard on N-Ba plane</td>
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<tr>
<td>H; Maximal cranial height</td>
<td>156.1</td>
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<td>153.5</td>
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<tr>
<td>L; Maximal cranial major diameter</td>
<td>195.7</td>
<td>167</td>
<td>185</td>
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<tr>
<td>Ha; Anterior cranial height in the N region</td>
<td>105.1</td>
<td>109.5</td>
<td>111.5</td>
</tr>
<tr>
<td>HP; Posterior cranial height in the Ba region</td>
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<tr>
<td>Frontal image</td>
<td></td>
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<tr>
<td>W; Maximal cranial width</td>
<td>168.3</td>
<td>164.5</td>
<td>163.5</td>
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<td>Cranial indices</td>
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<tr>
<td>H/L</td>
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<td>H/W</td>
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<td>W/L</td>
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<td>Profile</td>
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<td>S-N; Cranial major diameter</td>
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<td>S-Ba; Major diameter of the posterior skull</td>
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<td>S-ER'; Sphenoidal major diameter in the anterior skull base</td>
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<td>S-SOS'; Sphenoidal major diameter in the posterior skull base</td>
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<td>26.5</td>
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<td>NSBa; Basicranial angle</td>
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<td>S-ER'/S-N; Sphenoidal ratio in the anterior skull base</td>
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<td>0.53</td>
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<td>C) Maxillary region</td>
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<tr>
<td>Profile</td>
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<td></td>
</tr>
<tr>
<td>N-ANS; Anterior maxillary height</td>
<td>61.9</td>
<td>61.5</td>
<td>63.5</td>
</tr>
<tr>
<td>S-PNS; Posterior maxillary height</td>
<td>55.6</td>
<td>51</td>
<td>52.5</td>
</tr>
<tr>
<td>ANS-PNS; Anteroposterior major diameter in the upper jaw</td>
<td>55.5</td>
<td>48.5</td>
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<td>S-PNS/N-ANS; Anteroposterior ratio of the maxillary height</td>
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<td>D) Orbital region</td>
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<td>Profile</td>
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<tr>
<td>SN-IOR; Protrusion of the orbital anterior margin</td>
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<td>Frontal image</td>
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<tr>
<td>BIOD; Interorbital bone distance</td>
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<td>29.5</td>
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<td>BIOD/W; Ratio of the interorbital bone distance to the cranial width</td>
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<td>E) Mandibular region</td>
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<tr>
<td>Profile</td>
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<tr>
<td>Ar-Go; Mandibular ramus height</td>
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<td>48.5</td>
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<tr>
<td>Go-Pog; Mandibular corpus length</td>
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<td>76.5</td>
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<td>Ar-Pog; Mandibular length</td>
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<td>110.5</td>
<td>113</td>
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<tr>
<td>Gonial angle</td>
<td>123.4</td>
<td>132</td>
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<tr>
<td>Mandibular plane-SN, SNB</td>
<td>79</td>
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<td>67.5</td>
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<tr>
<td>Ar-Go/Go-Pog; Ratio of the mandibular ramus height to the mandibular corpus length</td>
<td>0.73</td>
<td>0.571</td>
<td>0.642</td>
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</table>

Analysis by method of Susami et al. demonstrated that maximal cranial major diameter was 185mm in calvarial profile, and sphenoidal major diameter in anterior skull base (S-ER') was 28.5mm in bascranial profile, indicating enhancement of dolichofacial pattern. In maxillary profile, anterior maxillary height (N-ANS), posterior maxillary height (S-PNS), and anteroposterior major diameter in upper jaw (ANS-PNS) had markedly increased to 63.5mm, 52.5mm, and 51mm, respectively, and in mandibular profile, mandibular ramus height (Ar-Go) and mandibular length (Ar-Pog) had also increased to 48.5mm and 113mm, respectively, indicating increase in high angle.
Fig. 7 Patient 2: Re-examination
Countenance: growth of face was observed. In oral cavity, occlusion was angle class II div. 1.

Facial Growth of Sotos Syndrome

Motohashi et al. performed a cross-sectional analysis of the morphology in infants with Sotos syndrome using Bolton standards. The subjects consisted of 4 patients: 3 boys aged 2 years 3 months, 3 years 1 month, and 5 years 8 months, and 1 girl aged 2 years 9 months. The cranial length, height, and width, maxillary and mandibular lengths, mandibular angle, and interorbital distance were measured on lateral and frontal cephalometric radiographs, and slight maxillary and mandibular retrusion, a large gonial angle, increases in the N-S-Gn angle in the mental region, increases in the Ar-Go-Gn angle, and slight increases in the facial height were detected.

Welbury and Fletcher reported an 11-year-old female and a 16-year-old male with Sotos syndrome, and noted their values by cephalometric analysis. Anterior cranial base (Sella-
Nasion) exceeded the norm in one patient, SNA was decreased, and SNPog was average or small. Mandibular body length (Go-Pog) was large and Gonial angle was average or small.

Our patients had a large anterior cranial base and exhibited dolichocephaly, and small SNA and SNB values.

Allanson and Cole\textsuperscript{11} reported growth changes in facial appearance using anthropometric craniofacial measurements. With time, the normal process of facial change occurs. They superimposed their results on this normal Gestalt. The results, it’s shamed that, as the face lengthened, the dominance of the forehead diminished and the chin achieved greater prominence. The mandible became longer and more narrow inferiorly, square or pointed, but prognathism was rare.

In the present study, we performed longitudinal analysis of the morphology in adolescent patients with Sotos syndrome, and found that the vertical development of the maxillofacial plane observed in the infants continued. To treat disordered occlusion in Sotos syndrome, it is considered important to suppress the growth of facial height.

**Conclusions**

To evaluate morphological changes in growth in patients with cerebral gigantism, we examined the maxillofacial growth pattern in
early teens using cephalometrical radio-
graphs of 2 patients who had consulted the
Orthodontic Department of Suido-bashi Hos-
pital. (Patient 1 was 8 years 6 months old at
the first examination and 14 years 6 months
old at re-examination, and patient 2 was 14
years 6 months and 16 years 11 months old,
respectively.) Ricketts and Downs & North-
western analyses and analysis by the method
of Susami et al. were performed using radi-
ographs of the 2 patients.

In both patients, vertical growth of the
maxillofacial plane was noted, and in patient
2, it was markedly enlarged.

The results showed that facial height
increased in the patients, with cerebral gigan-
tism occurring during the peripubertal period,
together with changes in vertical growth. This
suggests that these are important factors for
orthodontic treatment.

References

1) Allanson JE, Cole TR (1988) Sotos syndrome:
evolution of facial phenotype subjective and
objective assessment. Am J Med Genet 79:
229–230.

2) Gorlin RJ, Cohen MM, Hennekam RCM
ed., pp. 408–409, Oxford University Press, New
York.

3) Masubuchi M, Sueishi K, Sakamoto T, Negishi
F, Yamaguchi H (2006) Orthodontic evalua-
tion of-eight case in Beckwith-Wiedemann

Roentgencephalometric analysis of cerebral
gigantism: Report of four patients. J Craniofac

5) Nakazawa O (1959) A method of assessing the

6) Naritomi K (2001) Cerebral gigantism,
Nippon Rinsho Suppl Ryoikibetsu Shokokun
Shirizu No.33 of Molformation Syndrome

7) Sotos JF, Dodge PR, Muihead D, Crawford J,
Dtalbot NB (1964) Cerebral gigantism in
childhood: A syndrome of excessively rapid
growth with acromelic features and a non-

8) Susami T, Terashima T, Lin T, Motohashi N,
Kuroda T (1986) Morphological evaluation
on craniofacial dysmorphology of Apert’s syn-
drome and Crouzon disease. Jpn Orthod Soc

gigantism (Sotos syndrome)— two case reports.