Orthodontic treatment of Class II Division 1 malocclusion in a patient with achondroplasia

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Dwarfism occurs in about 1 in 26,000 individuals. Achondroplasia is the most common form of dwarfism, and about 90% of cases are caused by new mutations.[1] It is an autosomal dominant trait that affects males and females equally. Recent biochemical investigations indicate that the congenital defect factor receptor $FGFR^3$ is responsible for achondroplasia. It appears that recurrent mutations of a single amino acid in the transmembrane domain of the $FGFR^3$ protein is the culprit.[3] Manifestations of achondroplasia include short stature and craniofacial malformations,[2] including a short cranial base with early spheno-occipital closure and megalocephaly. Failure of normal cartilage proliferation leads not only to characteristically short arms and legs, but also to underdevelopment of the midface because the upper jaw is not translated forward by normal lengthening of the cranial base.[4] Cephalometric characteristics of achondroplasia include an enlarged calvaria with hydrocephaly and frontal bossing, short posterior cranial base, retrognathic maxilla, normal mandible, and protrusive maxillary incisors.[5,6]

The patient in this report had maxillary skeletal retrognathia and severe maxillary dental protrusion. This case was treated orthodontically and has remained stable during a 2-year retention period.

Case report

The patient, a 10-year-old Japanese girl with achondroplasia, was 109 cm tall at the initial visit. She was diagnosed as achondroplastic at birth and had been receiving growth hormone treatment three times per day for 2 years at the Department of Pediatrics, Tokushima University Medical Hospital. Her chief complaint was maxillary protrusion. She had a history of tongue thrusting and lower lip biting. Her dental history was unremarkable, and she appeared to be well adjusted, healthy, and intelligent.

Typical of achondroplasia, the patient exhibited a concave soft-tissue profile with a bulging forehead and a retrognathic maxilla. The nose was saddle-shaped because of lack of development of the nasomaxillary complex. There were no apparent major asymmetries, judging from both lateral and frontal photographs. The maxillary incisors were protrusive, causing the upper lip to protrude (Figure 1A-C).

A Class II Division 1 malocclusion was noted, with 8 mm overjet and 4 mm overbite in habitual occlusion. Minor irregularity was present in the mandibular premolar segment. The maxillary dental midline was deviated to the patient's left by 2 mm. No signs or symptoms of temporomandibular dysfunction were seen. The patient demonstrated a tongue-thrust swallowing pattern (Figure 2A-C).

Mounted cast analysis showed a Class II malocclusion with a 2 mm anteroposterior discrep-

Figure 1A-C
Pretreatment facial views

Figure 2A-C
Pretreatment intraoral views

Figure 3A-B
Lateral cephalometric and panoramic radiographs

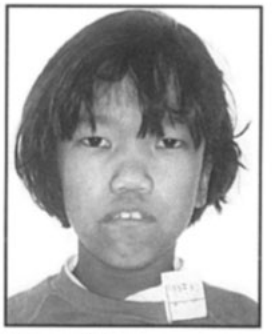

Figure 1A

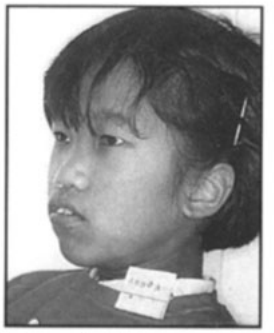

Figure 1B

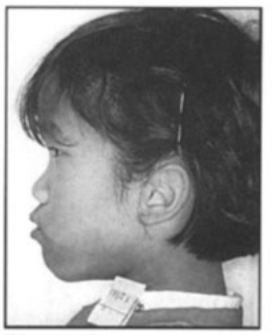

Figure 1C

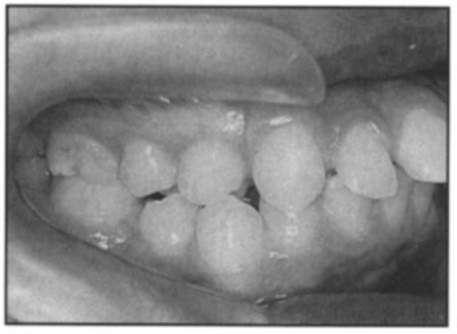

Figure 2A

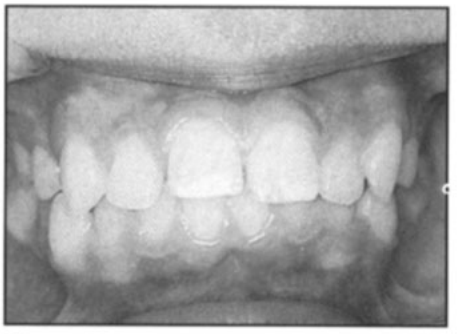

Figure 2B

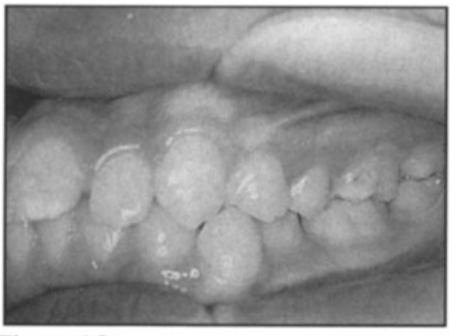

Figure 2C

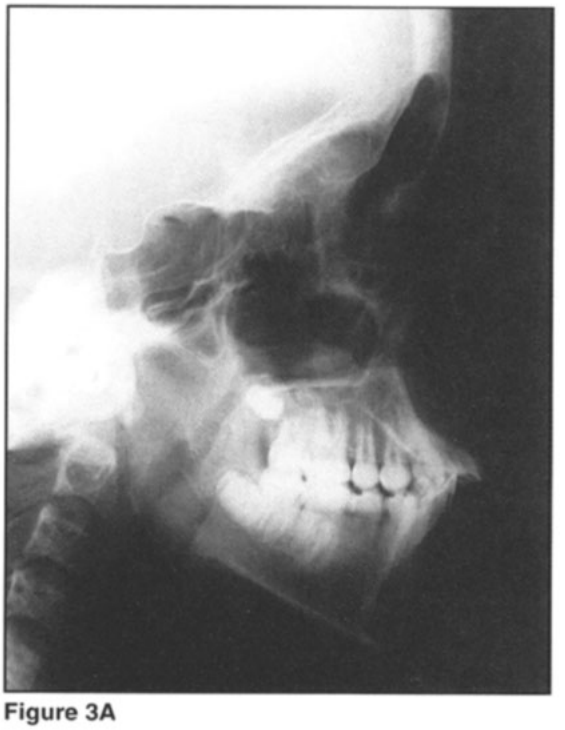

Figure 3A

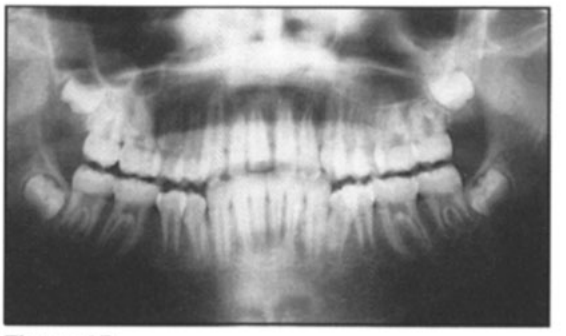

Figure 3B

ancy in the molar relationship. Mesiodistal crown diameters were larger for the maxillary premolars and first molars, and for the mandibular incisors, canines, premolars, and first molars, compared with Japanese norms.[7] Upper arch length was greater than normal.[8]

The panoramic radiograph revealed the presence of four third molar buds. There was a well-defined radiopaque odontoma at the apex of the left maxillary central incisor. The morphology of both mandibular condyles was unremarkable (Figure 3A-B).

Cephalometric analysis is presented in Table 1.

Skeletally, S-N (length of anterior cranial base) was more than 4 SD, indicating enlargement of the head. The saddle angle and S'-Ar' (posterior cranial base) were -2 SD. The SNA angle was -1 SD, indicating a retrognathic maxilla. The SNB angle was normal, ANB was -0.3°, and FMA was within 1 SD.

Dentally, U1-SN angle was greater than +6 SD, indicating proclination of the maxillary incisors to the apical base. IMPA was within 1 SD, and FMIA was 49.8°.

This analysis revealed a large head, a short cranial base, a retrognathic maxilla, a normal mandible, and protrusive maxillary incisors, thus vividly demonstrating the salient characteristics of achondroplasia.

Dentally, the patient had a Class II Division 1 malocclusion with maxillary incisor proclination.

Table 1
Summary of cephalometric analysis. S'-Ar': The linear difference between points S and Ar when projected on Frankfort plane

Measurement	Pretreatment 10 y 0 m	Mean Japan 12 y	Posttreatment 13 y 10 m	2 years posttreatment 15 y 10 m	Mean Japan 18 y
Facial A	78.8	83.1	79.9	83.2	84.6
Convexity	1.0	9.5	-5.1	-7.9	6.6
SNA	76.2	80.5	78.6	79.2	82.1
SNB	76.5	76.2	80.6	82.5	78.5
ANB	-0.3	4.3	-2.0	-3.3	3.6
Gonial A	123.4	128.3	120.7	118.8	121.2
FMA	35.1	32.4	32.5	29.2	28.6
IMPA	95.1	94.1	88.4	90.3	96.2
FMIA	49.8	53.5	59.1	60.5	55.2
U1 to SN	127.3	103.6	108.9	117.1	103.8

Skeletally, however, the patient had a Class III malocclusion with maxillary underdevelopment due to achondroplasia.

Treatment

The objectives of treatment were to achieve a Class I molar relationship, establish ideal overjet, correct the midline, and eliminate the tongue-thrust habit. A palatal holding appliance was placed and occipital headgear was fitted, followed by maxillary and mandibular first premolar extraction.

The steps of treatment were as follows:

1. Alignment and leveling
2. Maxillary and mandibular canine retraction
3. Maxillary and mandibular incisor retraction
4. Class II correction with Class II elastics.

Myofunctional therapy was encouraged to help correct the tongue thrust. After 41 months of edgewise appliance therapy, active treatment was completed and the patient was retained with maxillary and mandibular wraparound retainers. The odontoma at the apex of the maxillary left central incisor was extirpated, as were the four third molars.

Results

Total treatment time was 41 months due to poor cooperation by the patient. The lateral view (Figure 4C) shows a concave profile. However, the concavity seems to have remained relatively unchanged. Class I molar and canine relationships with acceptable overbite and overjet were achieved, and the pretreatment maxillary midline deviation to the left was corrected (Figure 5).

Cephalometric analysis at retention (13 years 10 months) showed that the ANB angle was reduced from -0.3° to -2.0° by retraction of the maxillary incisors and mandibular counterclockwise rotation from Class II elastics (Figure 7). On the basis of the changes, mandibular length increased more than maxillary length. The superimposition showed that approximately 10 mm of mandibular growth was expressed entirely as forward movement of the chin, while the temporomandibular joint remained in the same anteroposterior position relative to cranial base. Maxillary superimposition showed that the maxilla grew downward and the incisors became more upright and were retracted. Mandibular superimposition revealed that the mandible grew forward and downward, the incisors were retracted, and the first molars erupted normally. The face became slightly more concave as the angle of convexity increased (Figure 7).

Final evaluation

In general, cephalometric changes during treatment were favorable. These are summarized in Table 1.

At age 15 years 10 months, the chin-to-forehead relationship remained relatively stable and was slightly posterior to -7.9° (mean, +7.6°), and the angle of facial convexity became slightly more concave. All functional movements of the mandible were without limitation and without symptoms. Dentally, the maxillary and the mandibular

Figure 4A-C
Posttreatment facial views

Figure 5A-C
Posttreatment intraoral views

Figure 6A-B
Lateral panoramic and cephalometric radiographs

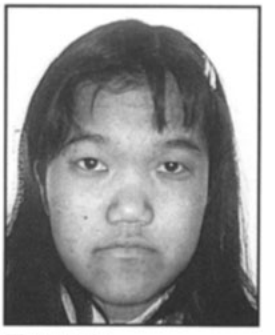

Figure 4A

Figure 4B

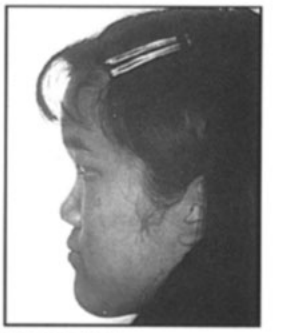

Figure 4C

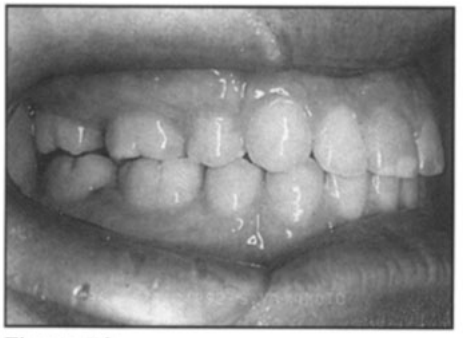

Figure 5A

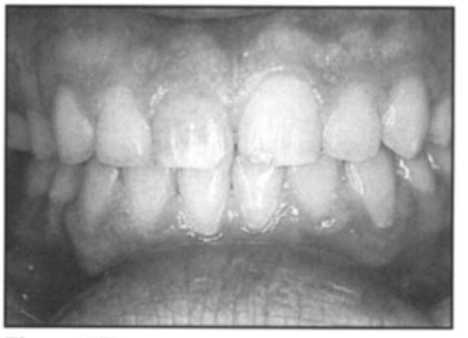

Figure 5B

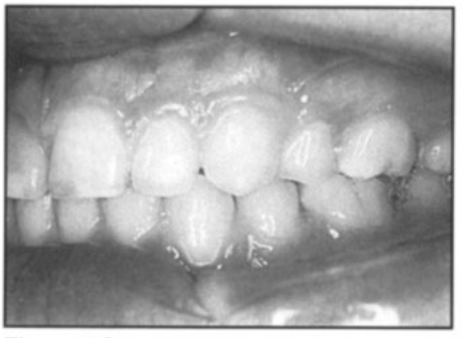

Figure 5C

Figure 7
Superimposed cephalometric tracings. The mandible grew in a forward and downward direction during treatment

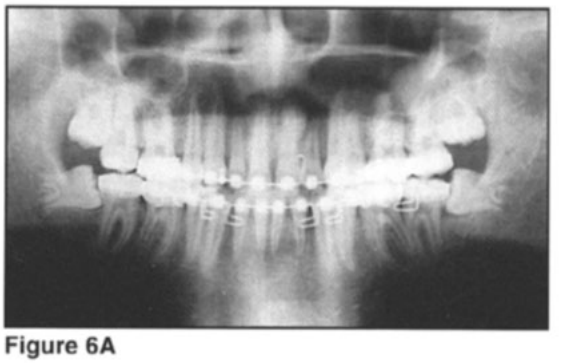

Figure 6A

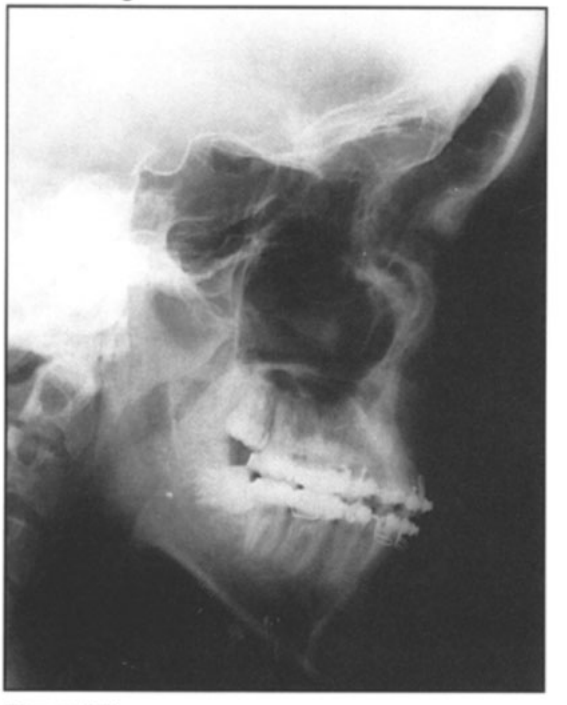

Figure 6B

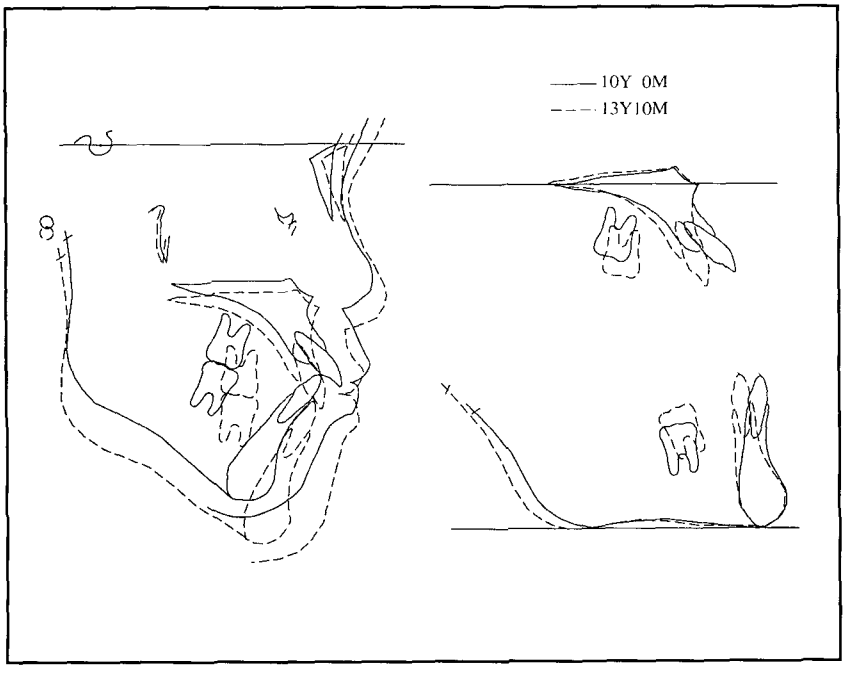

Figure 7

incisors were proclined, from 108.9° to 117.1° to the SN plane, and from 88.4° to 90.3° to the mandibular plane, respectively (Figures 8-11).

Discussion

The present case showed typical cephalometric characteristics of achondroplasia. Dentally, however, the patient displayed severe maxillary protrusion with lower lip biting and tongue thrusting. Therefore, the treatment plan included

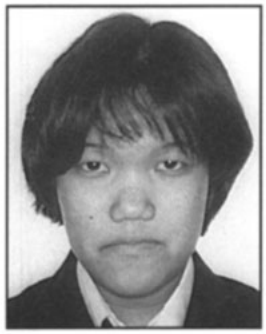

Figure 8A

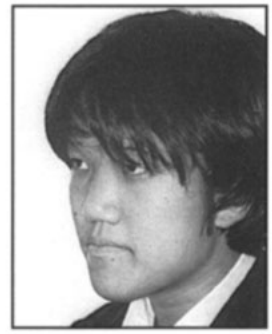

Figure 8B

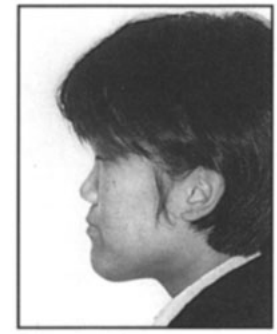

Figure 8C

Figure 8A-C
Facial views 2 years postretention

Figure 9A-C
Intraoral views 2 years postretention

Figure 10A-B
Lateral cephalometric and panoramic radiographs

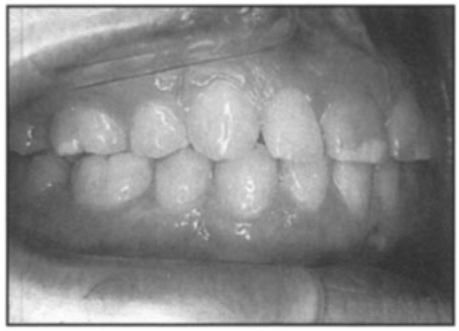

Figure 9A

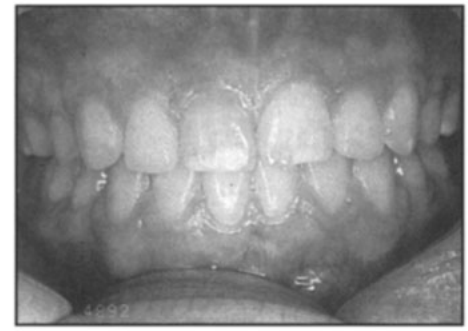

Figure 9B

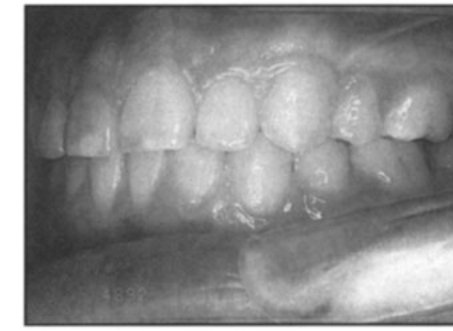

Figure 9C

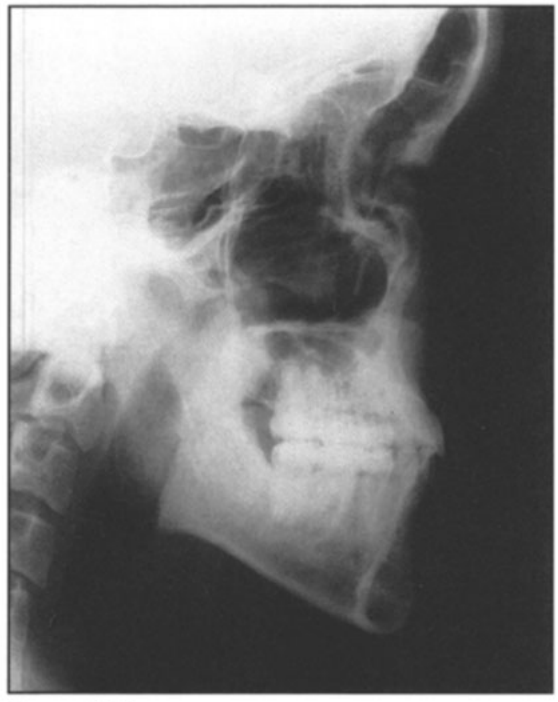

Figure 10A

premolar extractions to correct the protrusion. As a result, the ANB angle after treatment was reduced from its pretreatment value. At the time this manuscript was prepared, the patient was 15 years 10 months old and 119 cm tall, and growth was likely finished.[9] Tongue posture had improved, and occlusion had remained very stable during the 2-year retention period. No signs or symptoms of temporomandibular dysfunction were observed.

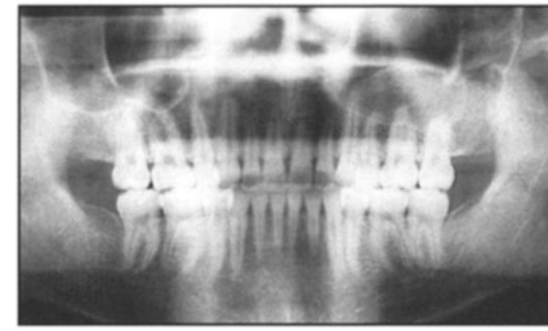

Figure 10B

Figure 11
Superimposed cephalometric tracings. Note the proclined maxillary and mandibular incisors

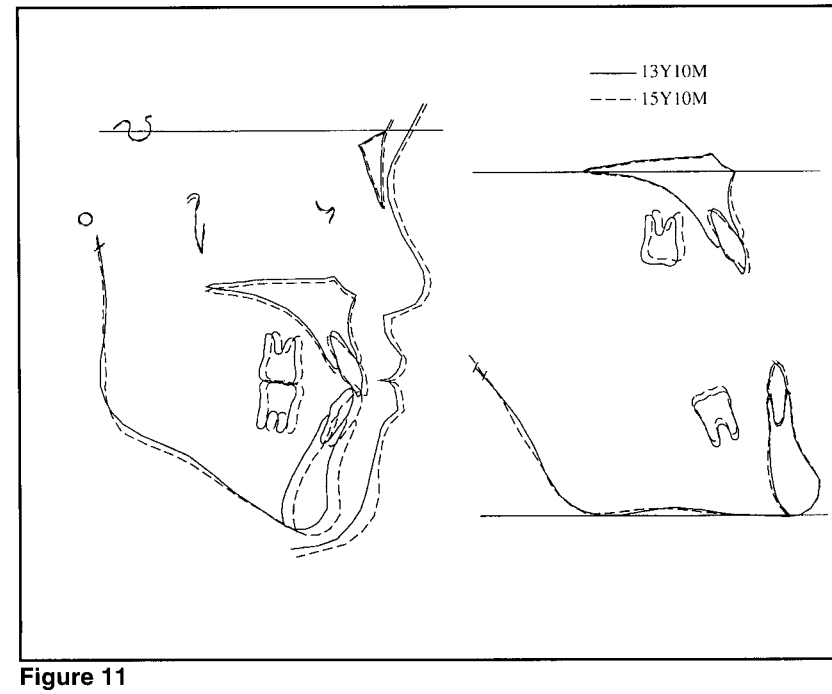

Figure 11

Some patients with achondroplasia are reported to require surgical treatment because of a discrepancy between the anteroposterior positioning of the upper and lower midface.[10,11] The clinical manifestation of this disorder that is of concern to orthodontists is the effect on the cranial base, which is characteristically small. Yet the cranial vault continues to grow, compensating for the developing brain. Treatment modalities may be limited since "growth" can't be used in the normal way.[12] Orthodontic treatment in this case was successful for the following reasons:

1. Maxillary growth was not inhibited by orthodontic maxillary protrusion
2. The skeletal deformity was not severe between the maxilla and mandible (ANB angle - 0.3°
3. Depth of the maxilla and morphology of the mandible were standard.

To date, there has been no medical treatment to improve height in constitutional bone disorders, such as achondroplasia and hypochondroplasia. However, patients with osteochondrodysplasias can be treated with growth hormones.[13] This patient received growth hormone treatment three times per day from 8 to 12 years. However, from examination of her overall height, sitting height, and rate of growth, the therapy did not accelerate her growth. Thus, no significant dose-response relationship could be demonstrated regarding the effect of the growth hormone treatment. Recently, in orthopedics, lower limb lengthening has been performed in dysharmonic hyposomia forms, such as achondroplasia and hypochondroplasia.[14] The aim is to increase the overall height and establish a better proportion between the trunk and the lower limbs. This patient received such surgical correction, which was used to lengthen both left and right tibias. As a goal, the lengthening should eventually increase her lower limbs a total of 15 to 20 cm.

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