



## Orthodontic and Surgical Treatment of a Patient with Apert Syndrome

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### ABSTRACT

The aim of this case report was to present the combined orthodontic and surgical treatment of a patient with Apert syndrome in an adult stage. A 15 years old male patient with Apert syndrome was concerned about the appearance of his face and malocclusion. His profile was concave with a retruded maxilla and prominent lower lip. He had an Angle class I molar relationship with a 9.5 mm anterior open bite. The amount of crowding was 20.4 mm in the maxilla and 6 mm in the mandible. Cephalometric analysis revealed a skeletal Class III relationship due to maxillary hypoplasia with a dolichofacial growth pattern. Orthodontic treatment and orthognathic surgery were planned for the patient. After 45 months of presurgical orthodontics, the patient underwent two surgeries sequentially. The first surgery was performed to advance the maxilla and the second surgery was performed to correct the mandibular rotation and increase the overbite at the time of removing halo device. The amount of maxillary advancement was 8 mm. Mandibula was moved 1.5 mm anteriorly and rotated  $1^{\circ}$  to  $1.5^{\circ}$  (SNB and facial depth) in a counterclockwise direction. After a relatively long treatment, an esthetically pleasing and functional occlusion and correction of the skeletal problem was achieved in this adult case.

**Keywords:** Apert syndrome, Maxillary hypoplasia, Orthognathic surgery.

**How to cite this article:** Kaya D, Taner T, Aksu M, Keser El, Tuncbilek G, Mavili ME. Orthodontic and Surgical Treatment of a Patient with Apert Syndrome. *J Contemp Dent Pract* 2012; 13(5):729-734.

**Source of support:** Nil

**Conflict of interest:** None declared

### INTRODUCTION

Apert syndrome is a rare genetic disorder characterized by premature fusion in cranial sutures, craniofacial anomalies and symmetric syndactyly of hands and feet.<sup>1</sup> On the face of a patient with Apert syndrome, high forehead, low-set ears, ocular proptosis, exophthalmos, hypertelorism, depressed nasal bridge and crossbow-shaped lips with prominent lower lip are observed.<sup>2,3</sup> The midface is retruded

with an increased total facial height.<sup>4</sup> The maxilla is hypoplastic<sup>5</sup> with an anteriorly tipped up palatal plane.<sup>2</sup> There are bulbous lateral swellings in the palate, suggesting a pseudocleft palate.<sup>6</sup> The mandible is smaller than normal and retrognathic<sup>2</sup> and a skeletal Class III malocclusion is observed. The most typical dental characteristics are an excessive anterior open bite, a severe crowding in the maxilla, a constricted and high maxillary arch, supernumerary teeth, delayed dentition, ectopic eruption and impacted teeth.<sup>3,5,6</sup> The crowding can be observed in the mandible but to a lesser degree.

The conventional treatment of the patients with Apert syndrome includes orthodontic treatment and orthognathic surgery. Rigid external distraction (RED) is one of the surgical treatment options for these patients.<sup>7-9</sup> Its feasibility in craniosynostotic syndromes has become widespread in recent years. Wakae et al<sup>10</sup> reported a functional and an esthetic treatment result with the RED system in a patient with Apert syndrome. In some instances, RED can be combined with conventional orthognathic surgery due to treatment need in the mandible or the directional movement limitations of the RED system.<sup>11</sup> In a case study, one case (18.6 years old) treated with maxillary distraction osteogenesis and bilateral sagittal split osteotomy in the mandible was reported.<sup>12</sup>

The aim of this case report is to present the combined orthodontic and surgical treatment of a patient with Apert syndrome in an adult stage.

### CASE REPORT

A 15-year-old male patient with Apert syndrome referred to our clinic due to the appearance of his face and malocclusion. He had typical clinical characteristics of the syndrome, such as high forehead, hypertelorism, exophthalmos, proptosis, midface hypoplasia, depressed

nasal bridge, low-set ears and syndactyly of feed and hand. He had fronto-orbital advancement at the age of 11 and several operations for syndactyly of hands. The profile was concave with a retruded maxilla and prominent lower lip. He had limited mouth opening (approximately 25 mm). The oral hygiene was poor due to the failure to use his hands. He had an Angle Class I molar relationship with a 9.5 mm anterior open bite. Maxillary midline was deviated 1.5 mm to the left side. Maxillary arch was constricted, a high palatal vault accompanying with pseudocleft palate was observed. Mandibular right and left second premolars did not erupt due to crowding (Figs 1A to G). The amount of crowding was 20.4 mm in the maxilla and 6 mm in the mandible.

Radiographic evaluation revealed that all third molars and maxillary right canine except for the mandibular left third molar were impacted. A supernumerary molar on the left side of the maxilla was observed (Fig. 2).

Cephalometric analysis revealed that he had a skeletal Class III relationship due to maxillary hypoplasia with a dolichofacial growth pattern. Maxillary incisors were labially inclined while mandibular incisors were normally inclined (Table 1, Fig. 3).

### TREATMENT PROGRESS

Orthodontic treatment and orthognathic surgery were planned for the patient. As the patient was 15 years old, the parents were informed and the treatment was delayed until the age of 18 years. At the start of treatment, an oral hygiene motivation was given. Maxillary right canine, maxillary and mandibular right third molars were extracted because of their unfavorable positions. Maxillary left first premolar and

second molar were also extracted because of arch length deficiency and root resorption respectively. The constricted maxillary arch was expanded with a Quad helix appliance. Maxillary left canine was distalized with a  $0.016 \times 0.016$  inch segmental retraction arch.  $0.018 \times 0.025$  inch Roth appliances were used to align the teeth in the dental arches. Presurgical orthodontic treatment lasted in 45 months (Figs 4A to F). After presurgical orthodontics, cephalometric and anteroposterior surgical treatment plans were made (Figs 5A to C) and the patient underwent two surgeries sequentially. The first surgery was Le Fort I osteotomy and distraction with RED I system to advance the hypoplastic maxilla. A rigid intraoral appliance was used for anchorage during distraction. On the fifth day of surgery, distraction was started at a rate of 1 mm/day. RED system was retained in place for 1 month (Fig. 6). After distraction, clockwise mandibular rotation and anterior open bite increased compared to the initial values. The second surgery was performed to correct the mandibular rotation and increase the overbite at the time of removing halo device.



Fig. 2: Pretreatment panoramic radiograph



Figs 1A to G: (A to F) Pretreatment extraoral and intraoral photographs, (G) hand-wrist radiograph of the patient

Clockwise rotation of the mandible was corrected after bilateral sagittal split osteotomy and advanced maxilla was fixed using plate and screws. Postsurgical orthodontic treatment lasted 13 months. Hawley type retainer was used for the upper and lower arches. Total treatment time was 60 months.

**RESULTS**

The combined orthodontic and surgical treatment approach largely improved the facial profile and dentoskeletal relationships of this patient with Apert syndrome. The amount of maxillary advancement was 8 mm. Mandible was moved 1.5 mm anteriorly and rotated 1° to 1.5° (SNB and facial depth) in a counterclockwise direction. At the end of the treatment, Class I canine relationship and Class II molar relationship were achieved on both sides. The maxillary and mandibular dental midlines were symmetric (Figs 7A to F). The post-treatment cephalometric measurements showed that SNA angle increased from 63° to 71° in combination with 2° of increase in SNB angle. ANB angle changed from -9° to -4°. FMA angle changed from a pretreatment value of 36.5° to a post-treatment value of 38°. The inclinations of maxillary and mandibular incisors were 105° and 87° for U1/SN and IMPA respectively. The initial anterior open bite, which was 9.5 mm, was changed to a vertical overlap of 1.5 mm. The positions of upper and lower

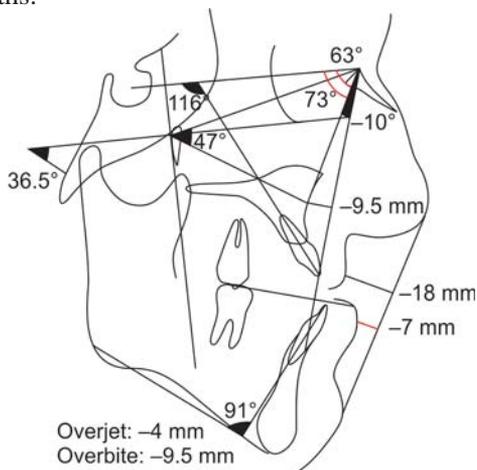
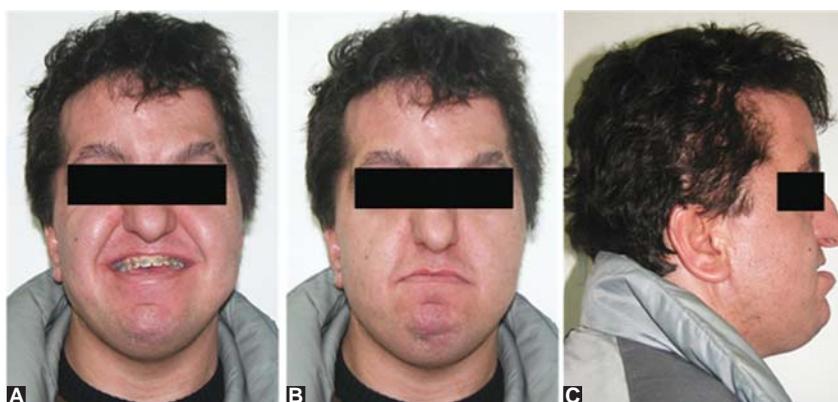
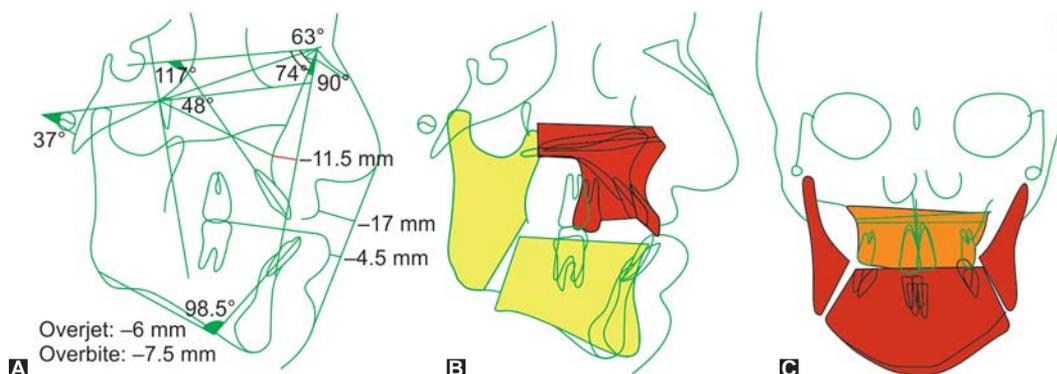


Fig. 3: Pretreatment cephalometric analysis



Figs 4A to F: Presurgical extraoral and intraoral photographs



Figs 5A to C: Presurgical cephalometric analysis (A) cephalometric (B) anteroposterior (C) surgical treatment plans

lips to E line became more favorable (– 11 and – 4 mm) than the initial positions (Table 1, Fig. 8A). The superimpositions of the pretreatment and post-treatment cephalometrics were shown in Figure 8B.

**DISCUSSION**

Surgical-orthodontic management of the dentoskeletal problem is the preferred treatment option in most of the Apert cases.<sup>13</sup> Before treatment, periodontal control is important in these patients because the crowded dentition and syndactyly of fingers reduce the ability to obtain good oral hygiene, causing greater dental plaque and periodontal problems.<sup>14,15</sup> The palatal bulbous swelling often leads to retention of food. This patient also had a poor oral hygiene

at the beginning of orthodontic treatment. He had a detailed periodontal treatment before treatment and an electrical toothbrush was recommended for improving his oral hygiene.

Maxillary constriction is a common feature in patients with Apert syndrome. A Quad Helix appliance was used to expand the maxilla in this patient. The need for tooth extraction in patients with Apert syndrome is also frequently reported in the literature.<sup>4,10,16,17</sup> However, the type of teeth extracted are different. Wakae et al<sup>10</sup> reported the extraction of maxillary right canine, maxillary left lateral incisor and maxillary left second premolar since they were in ectopic positions. Gugny<sup>16,17</sup> and Gugny and Vi Fine<sup>18</sup> extracted maxillary lateral incisors and premolars. Rynearson<sup>4</sup> extracted one premolar and several molars. Extracting severely impacted canines can shorten the presurgical orthodontic treatment time. In this patient, maxillary left first premolar due to crowding, maxillary right canine due to abnormal position and maxillary left second molar due to root resorption are extracted.

The complex dentoskeletal structure of this patient with Apert syndrome necessitated the use of RED technique and bilateral mandibular surgery to improve the severe craniofacial problem. Maxilla was advanced with the RED technique and bilateral sagittal split osteotomy was used to correct the clockwise rotation of the mandible and the occlusion. SNA and ANB angles increased 8° and 5° respectively.

It was reported that the treatment time for the patients with Apert syndrome was longer compared with the time



Fig. 6: Extraoral photograph after distraction with RED I system



Figs 7A to F: Post-treatment extraoral and intraoral photographs

required for nonsyndromic patients. The range was reported 33 to 58 months in the literature.<sup>4,12,16,17</sup> Similarly, the treatment time in this patient was quite long due to the severity of the problem and inadequate patient compliance.

The patient was very much satisfied with the treatment results. It can be stated without doubt that new facial appearance and functional occlusion made an important contribution in his quality of life.

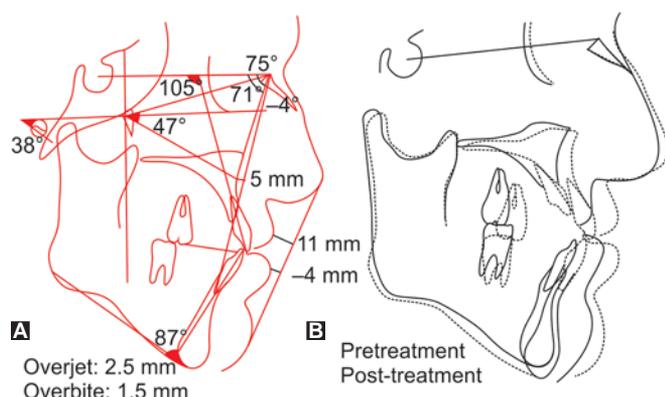
**CONCLUSION**

The treatment objectives for this Apert case included serial operations for sutural synostosis, syndactyly and correction of cranial bones in childhood, improvement of oral hygiene and periodontal status, elimination of dental crowding, establishing an esthetically pleasing and functional occlusion and correction of the skeletal problem by

orthognathic treatment. These treatment objectives were achieved by multidisciplinary approach in this adult case after a relatively long treatment time.

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**Figs 8A and B:** Post-treatment cephalometric analysis and superimposition of pretreatment and post-treatment cephalometric tracings

**Table 1:** Pretreatment, presurgical and post-treatment cephalometric values of the patient

Measurements	T0	T1	T3
SNA (°)	63	63	71
Maxillary height (°)	47	48	47
Maxillary depth (°)	63	61	70
Convexity (°)	-9.5	-11.5	-5
SNB (°)	73	74	75
Facial depth (°)	74	73.5	75
ANB (°)	-9	-9	-4
Overjet (mm)	-4	-6	2.5
Overbite (mm)	-9.5	-7.5	1.5
Lower facial height (°)	65.5	68	64.5
GoGnSN (°)	35.5	35.5	35
FMA (°)	36.5	37	38
U1-SN (°)	116	117	105
U1-FH	115	115.5	104
U1-NA (°)	51	52.5	33
U1-APog (°)	33	32.5	24.5
IMPA (°)	91	98.5	87
L1-NB (°)	19.5	28	17.5
L1-APog (°)	28	37	22
FMA (°)	52.5	44.5	55
Upper lip to E (mm)	-18	-17	-11
Lower lip to E (mm)	-7	-4.5	-4

T0: Pretreatment; T1: After first surgery; T2; Post-treatment

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