

## CASE REPORT

# Two-Phase Orthodontic Treatment in a Patient With Turner Syndrome: An Unusual Case of Deep Bite

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**Turner syndrome is caused by complete or partial absence of one X chromosome. These patients usually have a delay in growth and altered body proportions, causing sexual infantilism, short stature, delayed bone maturation, and variations in craniofacial morphology, among other systemic complications. The skeletal features associated with this syndrome include maxillary growth reduction with midface hypoplasia; mandibular micrognathia; high, narrow palate; V-shaped maxillary arch; and open bite. This case report shows a two-phase orthodontic treatment in a patient with Turner syndrome with a Class II malocclusion and severe deep bite, which is an unusual feature in patients with this disease. A conventional orthodontic treatment was performed, and after 20 months in retention the patient remains stable.**

KEY WORDS: *deep bite, orthodontic treatment, Turner syndrome, two-phase treatment*

Harry Turner (1939) identified the combination of short stature, lack of sexual development, webbed neck, and cubitus valgus found in seven women as Turner syndrome (TS). Ford et al. (1959) identified the genetic cause of TS as the absence of one X chromosome. Since then, different types of TS have been reported, the total absence of one X chromosome being the most severe form of the disease. Today a definitive diagnosis of this genetic variation is based on the chromosomal analysis of lymphocytes of peripheral blood or skin fibroblasts (Hall and Gilchrist, 1990).

Turner syndrome affects women, with a prevalence of 1 per 2500 girls born (Boue, 1938; Hook and Warburton, 1983; Hall and Gilchrist, 1990). The facial characteristics of TS are senile appearance, depressed lip angles, low-set ears, and multiple eye symptoms such as ptosis, strabismus, amblyopia, cataracts, and color blindness. Other clinical signs are short stature; primary amenorrhea; poorly developed genitalia with scarce pubic hair; pterygium colli; low hairline; broad chest and widely spaced nipples; narrowing of the aorta; multiple nevi; cubitus valgus; short fourth finger or toe; small, spoon-shaped nails; intestinal and renal problems; and feet and hand lymphedema (Gorlin, 1963; Lyons, 1968; Madléna et al., 1994).

Even though a delay in growth and altered body proportions are not pathognomonic clinical characteristics of TS, patients often present these features (Hall and Gilchrist, 1990; Vandewalle et al., 1993). This delay and the reduction in growth are due to the absence of a pubertal growth spurt, sexual infantilism, short stature, delayed bone age, and morphological craniofacial variations (Hall and Gilchrist, 1990; Rongen-Westerlaken et al., 1993; Vandewalle et al., 1993).

Chromosome alteration studies have shown that both the X and Y chromosomes promote dental growth, meaning that sexual chromosomes contain genes for dental development (Alvesalo, 1985). Among the different dental anomalies associated with TS are small teeth, simple crown morphology, thin enamel layer, short roots, and idiopathic root resorption (Fillipson, 1965; Motohashi, 1985; Varrela, 1990; Mayhall and Alvesalo, 1991; Midtbø and Halse, 1994). The skeletal characteristics associated with TS are reduction in maxillary growth with hypoplasia of the facial middle third; wide micrognathic mandible; anterior open bite; high, narrow palate; and V-shaped upper arch (Rongen-Westerlaken et al., 1993; Horowitz and Morishima, 1974). There have been also reports of patients with TS who have cleft lip and palate (Corona-Rivera et al., 2002).

There are only three reports of patients with TS treated with fixed orthodontic appliances and all of them had anterior open bites (Takeyama et al., 1990; Russell, 2001; Jivănescu et al., 2012). The purpose of this article is to report a two-phase orthodontic treatment in a patient with TS who presented a severe deep bite.

### CLINICAL FINDINGS

The patient was Colombian mestizo girl, 6 years 5 months old. The chief orthodontic complaint of the

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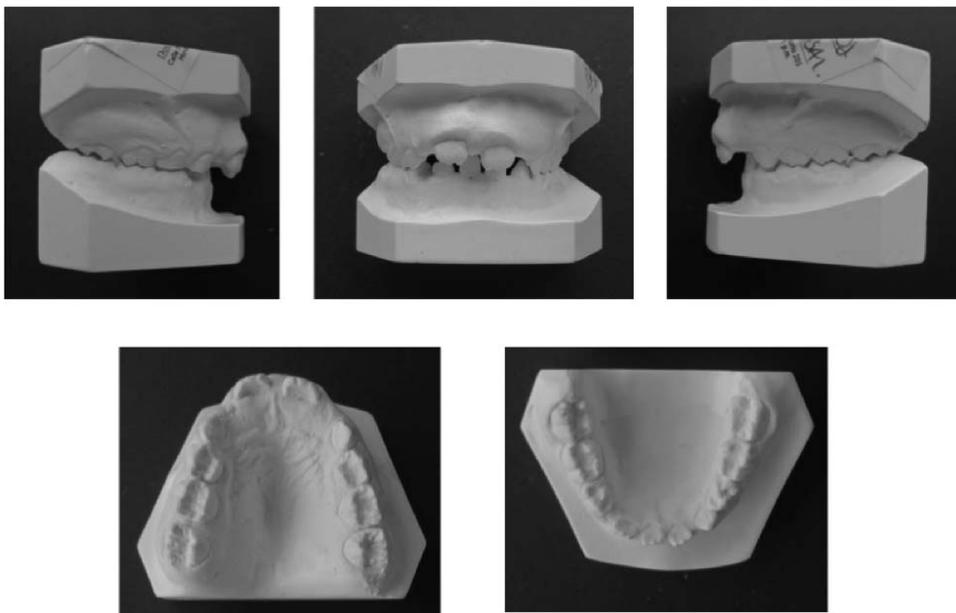


**FIGURE 1** Phase 1 pretreatment facial and intraoral photographs.

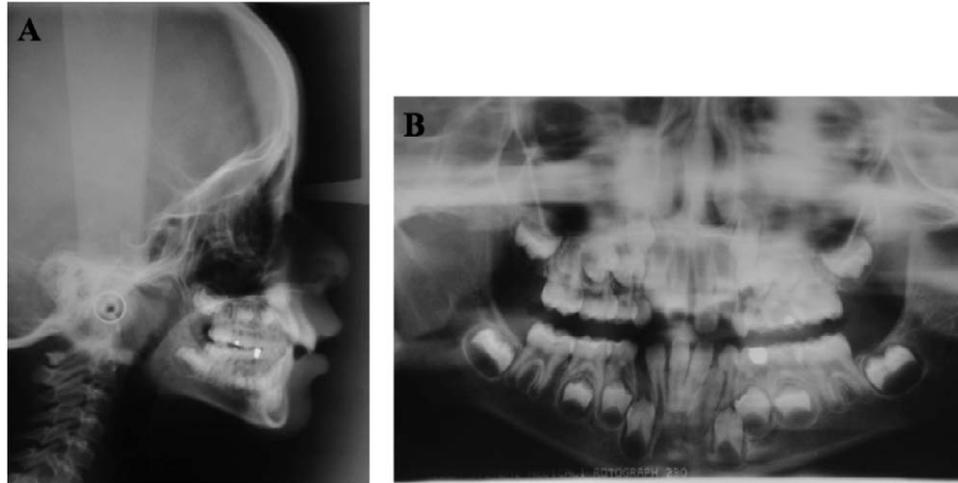
parents was that “she has a big clearance between upper and lower teeth.” Her clinical history included premature birth after 33 weeks, neonatal jaundice, severe oligohydramnios, patent oval foramen, and feet and hand lymphedema. Turner Syndrome was diagnosed by

means of karyotype where X chromosome monosomy was detected.

During her childhood the patient presented recurrent otitis media and was diagnosed with bilateral conductive hearing loss. She had hypothyroidism and a 3-year delay in



**FIGURE 2** Pretreatment dental casts.



**FIGURE 3** Phase 1 pretreatment radiographs. A: Lateral cephalogram. B: Panoramic radiograph.

her skeletal growth; thus, she was treated with growth hormones. Aortic coarctation was discarded, but the presence of a bicuspid aortic valve and mild tricuspid insufficiency were observed.

During the clinical evaluation were observed low-set ears, short and wide neck, and oval palate. She had a convex profile and a Class II malocclusion with an increased overjet and a deep bite (Figs. 1 and 2). Her cephalometric diagnosis (T0) was a skeletal Class II with micrognathia and retrognathia (Fig. 3; Table 1). She had a narrow airway space as a consequence of the maxillomandibular retrognathism, which may indicate an elevated risk of sleep apnea.

### TREATMENT OBJECTIVES

The following treatment objectives were established: (1) correct the Class II skeletal pattern; (2) improve the sagittal position of the mandible; (3) reduce the overjet and overbite; (4) guide the eruption of the permanent dentition; and (5) improve her facial aesthetics.

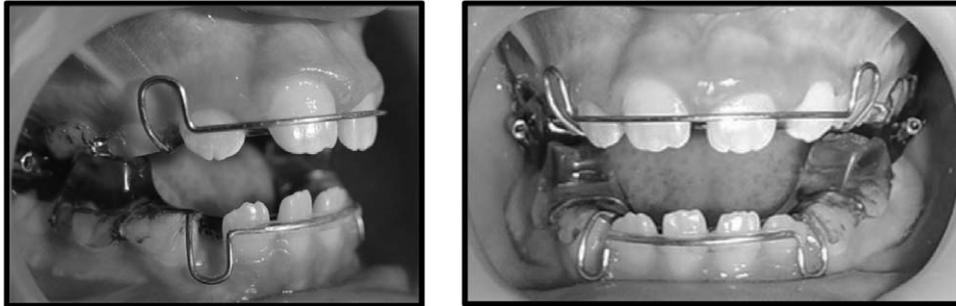
### TREATMENT ALTERNATIVES

There were some different treatment alternatives for the correction of the Class II malocclusion of the patient. The first treatment option considered was surgery when the

**TABLE 1** Cephalometric Values at Pretreatment Phase 1 (T0), Pretreatment Phase 2 (T1), and Posttreatment (T2)

| Measurements                               | T0   | T1  | T2  | T1-T0 | T2-T1 | T2-T0 |
|--|------|-----|-----|-------|-------|-------|
| SNA (°)                                    | 73   | 77  | 77  | 4     | 0     | 4     |
| SNB (°)                                    | 67   | 72  | 72  | 5     | 0     | 5     |
| ANB (°)*                                   | 10   | 6   | 6   | -4    | 0     | -4    |
| FH-MP (°)                                  | 26   | 24  | 28  | -2    | 4     | 2     |
| ANS-Me (mm)                                | 58   | 63  | 65  | 5     | 2     | 7     |
| Co-A (mm)                                  | 80   | 80  | 86  | 0     | 6     | 6     |
| Co-Gn                                      | 97   | 100 | 106 | 3     | 6     | 9     |
| U1-FH (°)                                  | 107  | 108 | 106 | 1     | -2    | -1    |
| U1-PP (°)                                  | 108  | 109 | 108 | 1     | -1    | 0     |
| U1-SN (°)                                  | 93.5 | 96  | 97  | 2.5   | 1     | 3.5   |
| Facial profile angle (G'-Sn-Pg)            | 160  | 163 | 165 | 3     | 2     | 5     |
| Nasal projection (Sn-P) (mm)               | 15   | 18  | 20  | 3     | 2     | 5     |
| Lower face height (Sn-Me') (mm)            | 65   | 66  | 68  | 1     | 2     | 3     |
| Lower face (Sn-Me'/G'-Me') (%)             | 56   | 53  | 51  | -3    | -2    | -5    |
| Chin projection (B'-Sn'Pg') (mm)           | 2    | 2   | 2   | 0     | 0     | 0     |
| Nasolabial angle (Col-Sn-ULA) (°)          | 98   | 101 | 105 | 3     | 4     | 7     |
| Upper lip length (Sn-ULI) (mm)             | 11   | 12  | 12  | 1     | 0     | 1     |
| Upper lip thickness (ULM-ULA) (mm)         | 12   | 13  | 15  | 1     | 2     | 3     |
| Maxillary sulcus contour (ULA-A'-Sn) (°)   | 160  | 157 | 156 | -3    | -1    | -4    |
| Upper lip protrusion (ULA-SnPg') (mm)      | 6    | 4   | 3   | -2    | -1    | -3    |
| Upper incisor exposure (StS-U1) (mm)       | 6    | 6   | 6   | 0     | 0     | 0     |
| Interlabial gap (StS-Stl) (mm)             | 8    | 1   | 3   | -7    | -2    | -5    |
| Lower lip-chin length (Stl-Me') (mm)       | 40   | 42  | 43  | 2     | 1     | 3     |
| Lower lip thickness (LLM-LLA) (mm)         | 11   | 12  | 13  | 1     | 1     | 2     |
| Mandibular sulcus contour (LLA-B'-Pg') (°) | 132  | 130 | 130 | -2    | 0     | -2    |
| Lower lip protrusion (LLA-SnPg') (mm)      | 4    | 5   | 4   | 1     | -1    | 0     |

\* Measurement with the Eastman correction.



**FIGURE 4** Phase 1 of treatment, orthopedics with Twin Block.

patient completes her skeletal growth, but due to possible complications during intubation, this option was dismissed. Another option was treating the patient with only the corrective phase with orthodontic fixed appliances, avoiding the orthopedic phase with the functional appliance, but the parents were concerned about the appearance of the girl and her self-confidence, in addition to the possible dentoalveolar trauma due to the protrusion of anterior teeth.

#### TREATMENT PROGRESS

It was decided that treatment should wait until the upper permanent lateral incisors erupted. The first phase of treatment was initiated when the patient was 7 years 4 months old. The patient used a Twin Block (Fig. 4) 24 hours a day for 15 months. The appliance had a 70° ramp with a lower buccal arch to control the inclination of the

lower incisors and an upper buccal arch that did not contact the upper incisors in order to reduce dentoalveolar compensation and correct the Class II skeletal pattern. The Twin Block was configured with larger posterior blocks, allowing anterior and upper rotation of the mandible and improving the mandibular Class II relationship. At the age of 8 years 7 months, an eruption guide and 4×2 orthodontic treatment were started to align and facilitate the eruption of upper permanent canines.

The second phase of treatment was initiated when the patient was 11 years 9 months and had permanent dentition. She had a convex profile and Class II malocclusion with increased overjet and deep bite (Fig. 5). Her cephalometric diagnosis (T1) was skeletal Class II with micrognathia and retrognathia, protrusion and vertical excess of the maxilla, acute nasolabial angle, lingual inclination and extrusion of the maxillary incisors, and labial inclination of the lower incisors (Fig. 6; Table 1).



**FIGURE 5** Phase 2 pretreatment photographs.

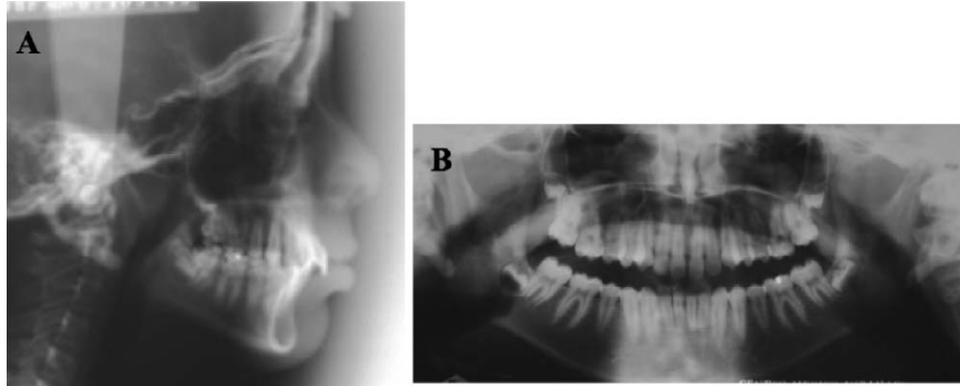


FIGURE 6 Phase 2 pretreatment radiographs. A: Lateral cephalogram. B: Panoramic radiograph.

It was decided to start the second phase of treatment with Orthos brackets (Ormco Corp., Orange, CA) slot  $0.022 \times 0.028$ . After leveling and alignment, the orthodontic treatment continued with intrusion of the upper and lower incisors (Fig. 7). A conventional arch sequence was followed. The patient used Class II elastics (3/16, 3.5 onz), and reverse curve of Spee arches were used from early stages of the treatment in the lower arch to manage vertical height. The finishing and detailing stage was done with titanium molybdenum alloy  $0.017 \times 0.025$  and  $0.019 \times 0.025$  arches. After 40 months of active orthodontic treatment, the Class II malocclusion was corrected and the deep bite, overjet, and overbite were improved (T2). Then it was decided to remove all her fixed appliances and

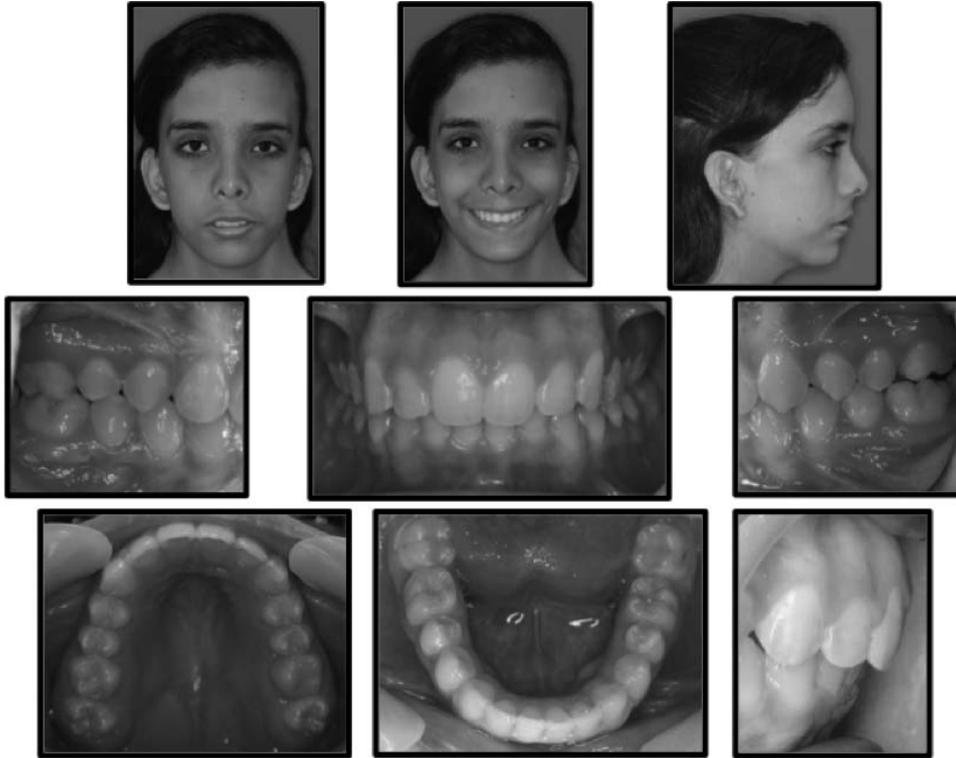
start retention with Hawley retainers (Figs. 8 and 9; Table 1). The patient was remitted to the periodontist for gingivectomy and to the oral surgeon for extraction of her impacted third molars (Fig. 10).

#### TREATMENT RESULTS

After phase 1 of treatment with the Twin Block appliance, the sagittal position of the mandible and the maxillomandibular relationship improved. The SNB angle increased  $5^\circ$  and ANB angle decreased  $4^\circ$ . Overjet decreased due to the proinclination of the lower incisors (Table 1).



FIGURE 7 Facial and intraoral photographs during phase 2.

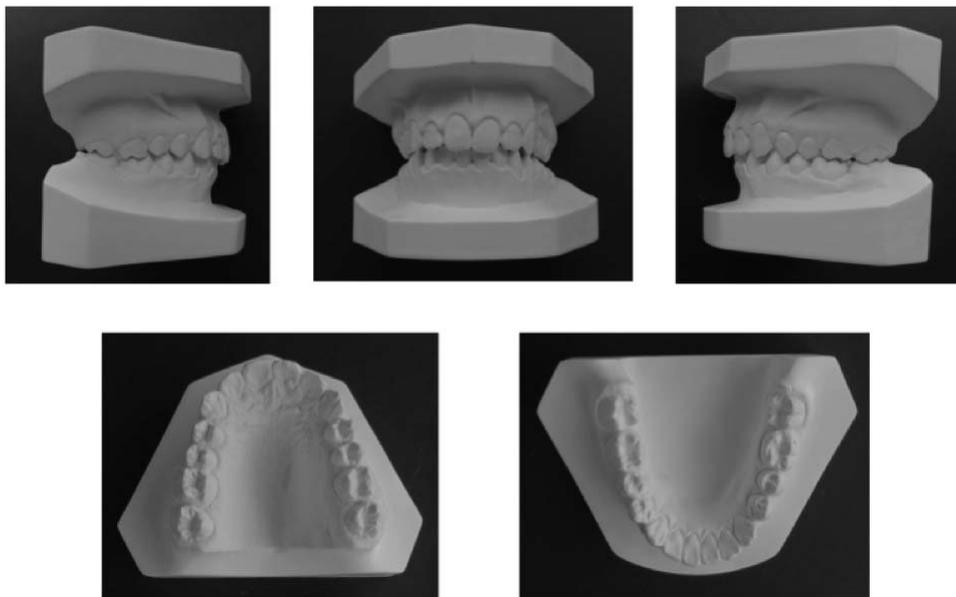


**FIGURE 8** Posttreatment facial and intraoral photographs.

During phase 2 of treatment with fixed appliances, the occlusal objectives were accomplished. The deep-bite was corrected, arches were widened, and overjet was fully corrected with the proinclination of the lower incisors, maintaining the inclination of the upper incisors. According to the soft tissue measurements

(Bergman et al., 2014) the facial profile angle, nasolabial angle, interlabial gap, and upper lip protrusion improved (Fig. 8; Table 1).

The patient was evaluated 20 months posttreatment; occlusal stability was observed and Class I occlusion was maintained, as was the correction of the deep bite (Fig. 11).



**FIGURE 9** Posttreatment dental casts.

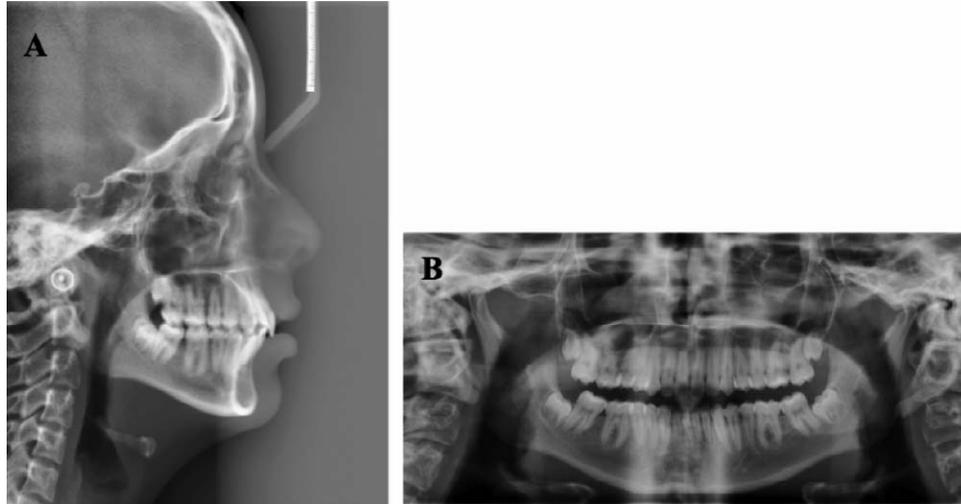


FIGURE 10 Posttreatment radiographs. A: Lateral cephalogram. B: Panoramic radiograph.

### DISCUSSION

In patients with syndromic conditions, it is of vital importance to have excellent compliance and commitment to fulfill all the objectives planned during the orthodontic treatment. This is even more important in two-phase treatments, which take longer to complete and sometimes are undertaken to avoid orthognathic surgery, where general anesthesia can result in important complications, especially in TS patients due to their anatomical and

physiological variations; a short neck and maxillary and mandibular hypoplasia could make it difficult to access the patient's airways (Maranhão, 2008). Also, because they have a shorter trachea and higher bifurcation point, these patients can be predisposed to bronchial intubation and accidental endotracheal extubation when the tracheal cannula is pulled from the airway (Maranhão, 2008).

In previous reports of patients with TS treated orthodontically (Takeyama et al., 1990; Russell, 2001; Jivănescu et al., 2012), all of them had open bites, which has been



FIGURE 11 Facial and intraoral photographs 20 months posttreatment.

associated with this pathology (Laine and Alvesalo, 1986; Rongen-Westerlaken et al., 1992; Midtbø and Halse, 1994; Szilágyi et al., 2000). However, this case report describes a malocclusion with a severe deep bite, which has been reported to have a low incidence in the TS population, varying from 3% to 31% (Laine and Alvesalo, 1986; Midtbø and Halse, 1996).

There are obvious advantages to treating Class II malocclusions with removable functional appliances prior to fixed appliance therapy. The orthopedic treatment of a Class II skeletal pattern can lead to improvement in orofacial function through muscle adaptation along with dental and skeletal changes (Harzer et al., 2010).

Although the ideal timing for orthopedic treatment for correcting mandibular retrognathia is after the onset of the pubertal growth spurt (Baccetti et al., 2000), and several studies have demonstrated that very early treatment involving two separate phases of therapy do not have any benefits (Tulloch et al., 2004; Dolce et al., 2007; O'Brien et al., 2009), this patient was treated in two phases to achieve a positive effect on her self-esteem (O'Brien et al., 2003).

The Twin Block functional appliance has several advantages, including the fact that it is well tolerated by patients (Harradine and Gale, 2000), is easy to repair, and is suitable to use in the permanent and mixed dentition. There are potential disadvantages such as the proinclination of the lower incisors (Jena et al., 2006) and development of a posterior open bite (Nayak et al., 2011). In this case report, the treatment objectives were achieved largely due to the good compliance by the patient. The patient's chief complaint was the increased overjet. Thus, by reducing the overjet with the functional appliance, the patient's confidence has improved, and the risk of sustaining trauma to the upper incisor was minimized (O'Brien et al., 2003).

This case shows that when the females with TS are treated with conventional orthodontics, good occlusal and facial balance can be achieved with the patients' compliance in both phases of the treatment, with good stability even after 20 months in retention.

## CONCLUSIONS

This two-phase treatment with Twin Blocks and fixed orthodontic appliances proved to be effective in the nonsurgical correction of a Class II malocclusion with a severe deep bite in a patient with TS.

The improvement in their facial aesthetics helps patients who have some type of syndrome compromise with their socialization processes and self-esteem.

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