

Surgery-First Technique for a Patient with Major β -Thalassemia

Milad Soleimani ^{a,b}, Mohammad Behnaz ^{b*}

^aDental Research Center, Research Institute of Dental Sciences, School of Dentistry, Shahid Beheshti University of Medical Sciences, Tehran, Iran; ^bDepartment of Orthodontics, School of Dentistry, Shahid Beheshti University of Medical Sciences, Tehran, Iran.

*Corresponding authors: Mohammad Behnaz, Department of Orthodontics, School of Dentistry, Shahid Beheshti University of Medical Sciences, Tehran, Iran, P.O. Box 19839. E-mail: behnaz1357@yahoo.com; Tel: +98-912 3278021

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Introduction: Thalassemic patients commonly present malocclusions, which require both orthognathic treatments and surgery. This study aimed to assess prioritizing the surgery over orthodontic treatment. **Case report:** Surgery-first method was performed to treat severe gummy smile and mild retrognathic mandible and prognathic maxilla in a 24-year-old Iranian girl with major β -thalassemia. Brackets and Ni-Ti wires were adjusted on all teeth to align their levels for one month. Then, the patient underwent the surgery, and the soft tissue was excised for Lefort dissection by two cutting lines, followed by osteotomy. Afterwards, down fracture and bilateral septoplasty were carried out, and excessive bones were removed. The surgery retruded the maxilla by 10 mm. **Results:** The 2-year follow-up revealed no overgrowth of the maxilla or malocclusion. **Conclusion:** Surgery-first method can be successfully used for treatment of malocclusions in patients with major thalassemia.

Keywords: Malocclusion; Orthodontics; Orthognathic; Surgery; Thalassemia

Introduction

Thalassemias are a group of inherited hematologic diseases caused by mutations in haemoglobin synthesis (1). Also, they are known as the most common genetic disorders worldwide (2) and are ranged from mild to severe, known as minor and major thalassemia, respectively. Accordingly, its mild forms are more frequently observed and can be managed, while severe forms are less common causing morbidity and mortality. There are two types of thalassemia as α thalassemia and β thalassemia, and these types refer to the deficiencies in the production of α and β globin proteins, which compose haemoglobin, respectively. This also leads to the production of abnormal red blood cells. A patient with thalassemia has the lowered levels of haemoglobin in their blood stream besides the low-quality haemoglobin (3).

The most severe cases of β thalassemia are usually diagnosed between the age of 6 months and 2 years old (4). The clinical signs associated with this type of thalassemia include severe and progressive anaemia, mild jaundice, pigment gallstones, cardiomyopathy, abdominal enlargement skeletal modifications resulting from the high blood iron levels and bone deformities.

Also, skeletal deformities including thinning of the cortical portions of the bones and pathologic fractures are common (5). Due to osteoporosis, the patients with thalassemia experience deformities in their jaw bones (e.g. enlargement of upper jaw or chipmunk face) and teeth. The lower jaw usually shows lesser enlargements because of its compact nature. Accordingly, this leads to malocclusion and speech, swallowing, and eating difficulties. Also, severe thalassemia often causes class II malocclusion (6)

Maxillofacial treatment for the patients with thalassemia involves orthodontic treatment, sometimes with surgery. In this regard, Einy *et al.*, in 2016 reported orthodontic treatment of a 10-year-old child with β thalassemia (7), who had severe aesthetic and functional disorders i.e., class II malocclusion. In the first five months, high pull Thurow maxillary orthopaedic splint was worn. By passing five months, a Twin block appliance was used for a seven-month period. Thereafter, the retention phase started that was lasted for 24 months. After these 36 months of treatment and retention, the patient had a stable class I molar relationship, and overjet and overbite have reduced. In another case, Gotte *et al.*, treated a patient with β thalassemia by applying the orthodontic and maxillofacial surgical corrective

treatment associated with an appropriate transfusion therapy (8). Their evaluations at different times showed that the therapeutic objectives had been achieved and maintained.

An alternative procedure is the “surgery-first” method. This procedure was proposed by Nagasaka *et al.*, (9) in 2009, this method proceeds with orthognathic surgery without presurgical orthodontic preparation and is followed by regular postoperative dental alignments. Although minor orthodontic movements are occasionally performed before surgery, the concept implies that most of the orthodontic treatment is performed postoperatively (10).

Compared with the traditional approach, surgery-first protocols lead to a significant decrease in total treatment time. This fact has a very positive influence on patients’ global satisfaction with treatment. The high orthodontic efficiency observed in surgery-first cases might respond to the combination of two factors. First, the starting point is the correction of the skeletal bases. In consequence, the complexity of orthodontic treatment is decreased, and soft tissue imbalances that might interfere with certain orthodontic movements are eliminated from the start. Second, tooth movement is accelerated owing to the increased postoperative metabolic turnover (11,12). This eventually leads to enhance psychosocial status of the patients, especially patients with severe conditions such as thalassemia.

This case study represented the importance of the surgery-first technique in dental and skeletal management (including surgical and orthodontic interventions) of a patient with major β thalassemia.

Case Report

A 24-year-old woman with chief complaint of severe gummy smile and mild retrognathic lower jaw and prognathic upper jaw was referred to the Oral and Maxillofacial Surgery Department of Taleghani Hospital, Tehran province, Iran. She had been diagnosed with major β thalassemia since her first year of birth and was then treated for over two years with removable orthodontic appliances since she was 10-year-old. Her history showed no other diseases except β thalassemia. She was treated with monthly transfusions. This study was conducted in terms of the fundamental principles of the Declaration of Helsinki (13).

The clinical and cephalometric evaluations (Figures 1 and 2 and Table 1) revealed a characteristic thalassemia facial appearance of skeletal class II relationship (maxillary protrusion and mild mandibular retrusion), grossly incompetent lips and gummy smile, trapped lower lip behaviour, and a severe convex facial profile with a normal TMJ function.



Figure 1. Preoperative clinical images



Figure 2. Preoperative cephalometric graphy



Table 1. Preoperative cephalometric measurements

Measurement	Definition	Initial
SNA	Angle that relates the maxilla to the cranial base (mean: $82\pm 3^\circ$), sella–nasion–A point angle	84
SNB	Angle that relates the mandible to the cranial base (mean: $80\pm 3^\circ$), sella–nasion–B point angle	77
ANB	A point, nasion, B point: indicating whether the skeletal relationship between the maxilla and mandible is a normal skeletal Class I (+2 degrees), a skeletal Class II (+4 degrees or more), or skeletal Class III (0 or negative) relationship. A point to B point angle	7
SN-Pog	The angle between sella, nasion and pogonion	78
SN Validity	The SN plane is valid if it has 6 to 8 degrees of divergent compared to the Frankfurt plane.	6
Max. depth	The angle formed by Frankfort plane and the N-A Angle	90
Mand. Depth	The distance between Ar and Pog	83
Facial depth	Formed by Nasion-Pogonion and Porion-Orbitale lines	84
Saddle angle	The angle between the anterior and posterior cranial base	117
Articular angle	The angle between sella, articular and gonion	158
Gonial angle	The angle between articular, gonion and menton	122
Occlusal plane	Occlusal plane through molars and premolars contact (functional plane)	18
Palatal plane	Formed by connecting ANS to PNS; used to measure the vertical tilt of maxilla	1
Inclination angle	The angle between palatal plane and perpendicular line from sella and soft tissue nasion	92
Wits	A simple method whereby the severity or degree of anteroposterior jaw dysplasia may be measured on a lateral cephalometric head film. The method entails drawing perpendiculars from points A and B on the maxilla and mandible, respectively, onto the occlusal plane.	+5
Interincisal angle	The angle between upper and lower incisors	119
Upper 1 to SN	The angle between long axis of lower incisor and SN plane	108
IMPA	The angle between long axis of lower incisor and mandibular plane angle	97
Jaraback index	Proportion of posterior height (S-Go) to anterior height (N-Me)	64%
Y-axis	Sella-Gnathion to Frankfurt Horizontal Plane	70



Also, medical evaluation, including chest radiography, complete blood count, blood smear, liver function tests, electrocardiogram, and thrombin and partial thromboplastin time were performed. Hematologic and cardiovascular consultations revealed the patient's corrected hematologic situation. The preoperative haemoglobin level of 9.4 g/dL was deemed sufficient by the haematologist to allow the surgical correction of the facial deformity.

In this study, to treat the deformity, we used "surgery-first" method proposed by Nagasaka *et al.*, (9) who have used this method to correct skeletal class III malocclusion. Their treatment plan was performed as follows: first, they bonded pre-adjusted brackets to all teeth, except the maxillary second molars. Then, model surgery was performed and a surgical splint with a lingual bar and ball end clasps were made to cover the posterior occlusal surfaces. Afterwards, bilateral sagittal split ramus osteotomy was performed to achieve the required mandibular setback. Orthodontic treatment started one month after the surgery. Using this method, the total treatment time reduced in comparison to the traditional orthognathic treatment, which includes pre-operative orthodontic preparation, surgery, and a relatively constant period of post-operative orthodontics.

In the present case, before the surgery, all teeth were checked for any possible infections. Then, we used brackets on all teeth and Ni-Ti wires to align their levels for one month, and

after that, passive rectangular wires were used. After thorough analyses of occlusal and skeletal relationships, the patient underwent general anaesthesia. Also, soft tissue was excised for Lefort dissection by two cutting lines as follows: one 7 mm higher than the root of the teeth and the second one 5 mm higher than the first line. Two Lefort cuts were performed as well as osteotomy. Afterward, down fracture was performed as well as bilateral septoplasty, and excessive bones were then removed. During the surgery, gentle dissection and conservative drawing surgical cut were performed to lessen the blood loss. Furthermore, five pack cell also reserved for the patient to be prepared for the possible blood transfusion during the surgery. In an optimal occlusion, maxilla was retruded 10 mm and fixed with four L-shaped plates. Finally, the surgical area was sutured with a 4-0 acrylic suture.

Post-surgery orthodontic treatment was performed using .014" stainless steel wires, followed by 0.18" stainless steel wires. At this stage, level of teeth was aligned and dental arch was then coordinated.

Results

Since the patient was not fully cooperative for using elastics, the outcome was not as ideal as expected. However, a 2-year follow-up revealed no overgrowth of the maxilla or malocclusion (Figures 3 and 4).



Figure 3. Postoperative clinical images



Figure 4. Postoperative cephalometric graphy



Discussion

In this study, we reported the successful treatment of a severe gummy smile and mild retrognathic mandible in a 24-year-old Iranian girl with and major β -thalassemia. The “surgery first” method was performed to treat the deformity. During the surgery, maxilla was retruded 10 mm. Also, a 2-year follow-up found no overgrowth of the maxilla or malocclusion which shows the success of our method.

Patients with severe dental and skeletal anomalies experience many psychological problems pertaining to their anomalies. It is believed that orthognathic surgery not only does correct the physical anomalies, but it also does help enhance psychosocial health (14,15). Furthermore, as skeletal problem (and therefore the esthetic concern) is corrected from the beginning, patients have better compliance with postoperative orthodontics and this is a powerful contributor to global satisfaction with treatment. Thus, performing surgery can have significant impacts on the patients’ lives.

It is important not only to determine the baseline hemoglobin level, but also to assess the percentage and types of hemoglobin the patient carries. HgF and HgA2 differ from normal HgA in that they have increased oxygen affinity, not readily releasing oxygen from the iron core. This can potentially compromise the patient postoperatively because proper oxygen distribution is required for optimal healing environment. In the present case, the constitution of our patient’s hemoglobin was 100% HgF.

Surgical management of thalassemia- induced facial deformities includes repositioning and contouring of the maxilla. Patients with severe deformity with a complete lack of cortical bone for bone fixation might not be amenable to such procedures. Surgical resection and recontouring in a patient with a thalassemia- induced facial deformity has been previously reported (16). Surgical resection to establish the proper alveolar height and width to receive a partial or complete removable prosthesis is possible and should be considered for extreme facial deformities.

The “surgery first” concept in orthognathic surgery was introduced by Nagasaka *et al.*, in 2009 (9). Accordingly, they reported the correction of a Class III skeletal malocclusion with mandibular setback surgery and subsequent orthodontic alignment with the aid of temporary anchorage devices. The patient underwent no previous orthodontic preparation. Due to their excellent clinical results and substantial reduction in total

treatment time, the investigators postulated that, this new treatment approach could become a standard procedure in future. By considering the number of patients requesting orthognathic surgery with primarily aesthetic concerns and time limitations for performing a long-term treatment, the “surgery first” approach could be represented as a reasonable, cost-effective method to manage skeletal malocclusion in the selected cases of thalassemia. However, to the best of our knowledge, there are few references to the use of the “surgery first” approach in bimaxillary orthognathic surgery in the published scientific reports.

Surgery-first procedure could be approached in two methods: “surgical-driven” and “orthodontic-driven”. The former corrects both the jaw and dental deformities via the surgical procedure (17); while, the latter corrects the jaw deformity by surgery and the dental deformity via orthodontics (18). We used orthodontic-driven method in this study. While surgical-driven method includes fewer steps and maybe more pleasing to patients, orthodontic-driven method may produce more favourable results as it involves orthodontic step.

The dentoalveolar class II relationships found in our patient, have been frequently reported. Pusaksrikit and Isarangkura also found class II malocclusion among the thalassemia major patients (especially in men) with no class III tendency noted, despite its high incidence of 20% among the Thai population (19). This has been attributed to the slower mandible growth among men as it was blocked by the excessive maxillary growth. However, in a follow-up study on 88 patients with thalassemia from the same population, Pusaksrikit and Isarangkura noted that, the patients had a tendency toward class I malocclusion with a maxillary anterior dentoalveolar protrusion. The inclination of the incisors in our patient represented dentoalveolar compensation mechanisms to prevent the increased overjet in such a marked maxillo-mandibular discrepancy.

We used “surgery first” method in this case, and gained 10 mm retrusion of maxilla. Notably, most case reports found in the literature have gained less than this amount (16,20,21). For instance, in a study by Mortazavi and Khojasteh (20), three cases with thalassemia (two of them with thalassemia intermedia) were surgically treated. In the patients with thalassemia intermedia, 7 to 8 mm repositioning was performed. Furthermore, this approach lowers the treatment time, which can be more pleasant for both the patient and practitioners. Another advantage of the “surgery first” approach compared to



the traditional surgical-orthodontic treatment includes the rapid profile improvement with subsequent immediate patient satisfaction. In addition, in the case of the skeletal Class III patients, the upper lip and tongue tone improvements occurring after mandibular setback increase the force on the incisors, improve the efficiency of incisor decompensation, and reduce the total treatment time.

Post-operative relapse in this patient was managed by elastics and orthodontic appliance. Although it is uncommon (22), generally, relapse within the immediate surgical period is caused by inadequate mobilization of the repositioned jaw. Other possible causes include bony interferences and instability not appreciated during the repositioning; condyles dislodged from the glenoid fossa at the time of fixation; and failure of the internal plate/screw fixation systems. Besides, patients with thalassemia have higher chances of relapse following further growth. To prevent this, it is suggested that the surgeon might consider omitting intermaxillary fixation initially and placing elastics two to three days after the procedure in a more relaxed setting. Additionally, an occlusal relapse can occur late, long after the initial surgery. Late relapse usually results from complex functional dental and muscular forces re-establishing equilibrium and remodeling of the facial skeleton. Long-term follow-up is important, and the management is targeted toward the causes (23). However, it should be considered that patients with thalassemia have thinner cortical plates and prolonged treatment with myofunctional appliance and fixed orthodontic treatment at younger ages can increase the chances of root resorption (24). Hence, the duration of fixed orthodontic treatment and maintenance should be as minimal as possible.

Conclusion

The surgery-first method can be a great alternative to conventional treatment plans for treatment of dentomaxillofacial malocclusions in patients with major thalassemia, as it decreases the complexity of orthodontic treatment and the total treatment time, and consequently increases the patient's satisfaction.

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Conflict of Interest: 'None declared'.

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