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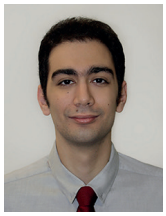

Maziar Farhadi

Rosana Farjaminejad, Samira Farjaminejad, Anand Marya and Abdolreza Jamilian

Surgical orthodontics in a patient with β-thalassaemia major. A case report

Abstract: Treating patients with prognathic maxillae is a multifaceted process, especially when dealing with thalassaemia complications. This case study addresses the issue of severe maxillary protrusion in a woman living with β-thalassaemia major who had presented with pronounced maxillary protrusion, lip incompetence and an everted upper lip. Following initial alignment and levelling using the 0.018-inch standard edgewise system, the patient underwent a Le Fort I osteotomy procedure. Segmental osteotomy was ruled out owing to the heightened risk of excessive bleeding. The outcomes demonstrated successful management of the thalassaemic patient through pre-surgical orthodontics, LeFort I osteotomy, and subsequent post-surgical orthodontics and periodontics. The patient exhibited improved facial aesthetics after an active treatment period of 4 years.

CPD/Clinical Relevance: Orthodontic treatment for patients with β-thalassaemia major and skeletal Class II malocclusion with prognathic maxillae requires a multidisciplinary approach, including Le Fort I osteotomy and careful management of elevated bleeding risks

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Thalassaemia is a family of hereditary genetic conditions that are common worldwide. They can be categorized into two primary divisions: α- and β-thalassaemia.[1] β-thalassaemia, alternatively referred to as Cooly's anaemia, is an inherited blood disorder marked by an abnormality in skull formation, resulting in a distinctive appearance in the affected individual.[2]

Clinically, β-thalassaemias can be categorized as thalassaemia trait, minima, minor, intermediate and major, indicating the level of anaemia present. This disorder primarily affects regions around the Mediterranean Sea, a significant portion of Central Africa, the Middle East, the Indian subcontinent, and southeast Asia, encompassing Indonesia, with occurrence rates ranging from 5% to 20%.[1] These haematological challenges contribute significantly to orthodontic complications, because the disorder is associated with extensive craniofacial changes as a result of bone marrow hyperplasia and altered bone metabolism, complicating standard orthodontic treatments.[3] Thalassaemia may be linked to skeletal facial deformities.[4]

Characteristic craniofacial attributes seen in individuals with β-thalassaemia major include a Class II skeletal arrangement, a reduced length of the cranial base, distinct vertical growth direction of the mandible, diminished mandible size, enlarged upper jaw, lips that turn outward, narrowed nasal passage, and heightened frontal facial dimensions.[5]

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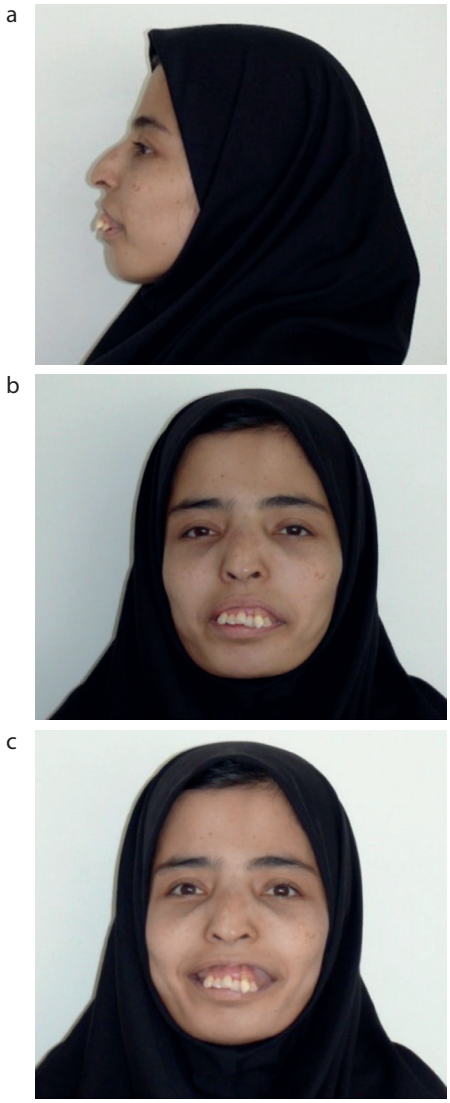


Figure 1. (a–c) Extra-oral pre-treatment photographs.

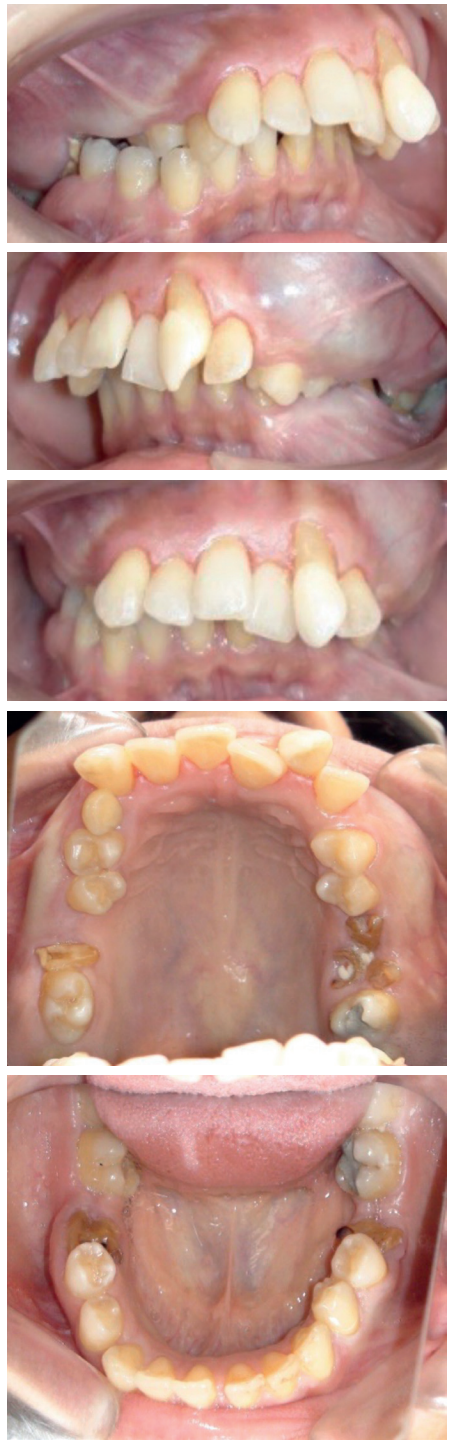


Figure 2. (a–e) Intra-oral pre-treatment photographs.

Individuals with thalassaemia often exhibit a distinctive craniofacial and jaw growth pattern, accompanied by prominently protruding lower incisors.[6] The arrangement of teeth demonstrates protrusion, widening, and spacing among the front upper teeth, along with noticeable prominence of the lower incisors.[7–9] These dental and skeletal anomalies necessitate specialized orthodontic approaches that accommodate the unique structural challenges and increased surgical risks, such as excessive bleeding and delayed healing, associated with the disease.[10]

Typically, following the removal of two premolars, a segmental osteotomy is performed to reposition and align the maxilla. However, owing to significant concerns about blood loss in these patients, the option of segmental osteotomy is typically deemed to be unsuitable. A comprehensive examination of the existing literature identified minimal case documentation involving Le Fort 1 osteotomy and orthodontic treatment in β-thalassaemia major cases. In this case report, the application of Le Fort I osteotomy and orthodontic treatment to reposition the upper jaw and address malocclusion in a patient with thalassaemia major is presented. This treatment approach aligns with evidence-based practices discussed in a Cochrane review that evaluated the effectiveness and safety of dental and orthodontic interventions for treating skeletal deformities and other complications in patients with thalassaemia.[11] The review highlighted the unique challenges faced in managing orthodontic and surgical interventions for thalassaemia patients, particularly those related to the increased risks of excessive bleeding and delayed healing. The successful use of Le Fort I osteotomy in this case supports the feasibility of combining orthodontic and surgical methods to enhance both aesthetics and function in individuals with complex medical conditions such as β-thalassaemia major.

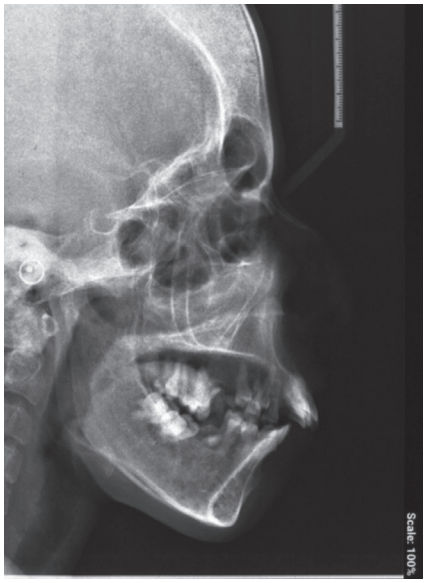
Case report

A 25-year-old woman diagnosed with β-thalassaemia major was referred to the orthodontic department. She presented with the main concern of a protruded upper jaw and forward positioning of her upper front teeth. At the time of her consultation, the patient had no concurrent cardiovascular, pancreatic or other medical issues. It was noted from her medical history that she had undergone spleen removal when she was 9 years old. On examining her facial structure, it was observed that she had a protruding and excessively enlarged upper jaw, with excess gingival display on smiling, incompetent lips, and an everted upper lip. Gingival recession was obvious on the upper left canine. There was also a lower lip trap (Figure 1).

Intra-oral examination revealed crowding of the upper incisors, a deep curve of Spee, increased overjet and overbite, and a severe Class II malocclusion. Although all third permanent molars were present, all four first permanent molars were unrestorable, leaving only remnants of the roots intact (Figure 2).

The pre-treatment lateral cephalometric and panoramic radiographs are shown in Figure 3. The patient's lateral cephalograms were traced before and after treatment. Tracing was carried out manually with a ruler and protractor. The measurement accuracy was checked by tracing each lateral cephalogram twice by the same

a



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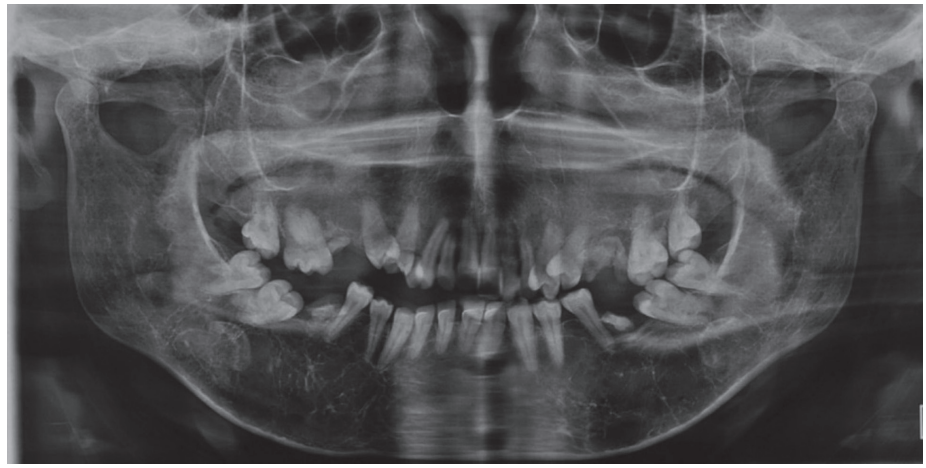

Figure 3. Pre-treatment lateral cephalometric and panoramic radiographs.

Landmarks	Pre-treatment
SNA	85°
SNB	74°
ANB	11°
GoGn/Sn	46°
Y-axis	70°
U1–Sn	105°
IMPA	109°
Interincisal angle	100°
Nasolabial angle	92°
Facial convexity (G'-Sn-Po')	22°

Table 1. Pre-treatment cephalometrics of the patient.

researcher. Intra-class correlation coefficients (ICCs) calculated for each variable showed the measurements to be consistent at 95%, indicating no random error (Table 1).

Treatment objectives

The treatment goals for this individual were to:

- Correct the maxillary protrusion by repositioning the upper jaw using Le Fort I osteotomy to reduce the overjet to a more clinically acceptable range.
- Improve dental occlusion by transitioning from a Class II to a more neutral occlusal relationship.
- Effectively manage the spacing issues through orthodontic alignment and preparation for potential restorative solutions to replace the missing teeth, ensuring a functional and aesthetic dental arch.

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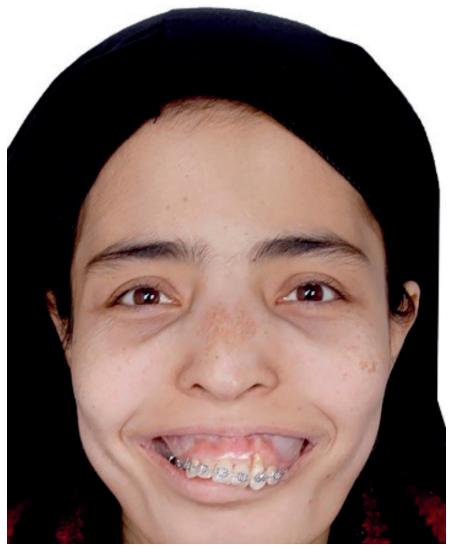

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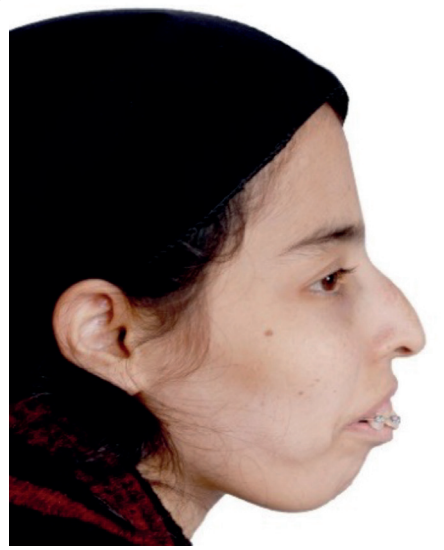

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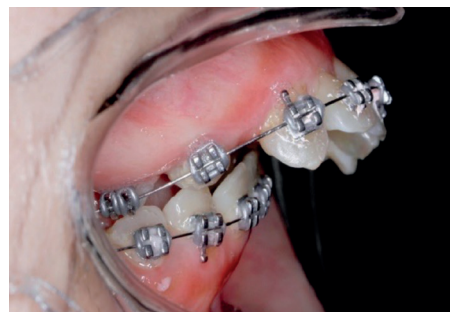

Figure 4. (a–c) Before surgery photographs.

Alternative treatment

The option of extracting four first permanent molars followed by segmental osteotomy was contemplated as an alternate therapeutic approach for the patient. However, this was considered inappropriate owing to the heightened bleeding risk it posed. As a less invasive alternative, the treatment team considered using dental implants to address the spacing. This approach aimed to minimize surgical risk, while effectively managing the the missing teeth.

Treatment progress

Before starting the orthodontic treatment, the remaining roots of the upper first molars and the lower right and left first molars were removed. After applying the 0.018 standard edgewise system for levelling and alignment, the overjet was augmented. The curve of Spee was flattened within the lower arch.

Subsequently, coordination was established between the upper and lower arches. Stainless steel archwires of dimensions 0.017"x0.025" were inserted into the brackets pre-operatively (Figures 4 and 5). At this juncture, the patient exhibited an overjet of 22 mm and was prepared for Le Fort 1 surgery.

In this case, a Le Fort I osteotomy was preferred over a segmental osteotomy despite the associated bleeding risks, based

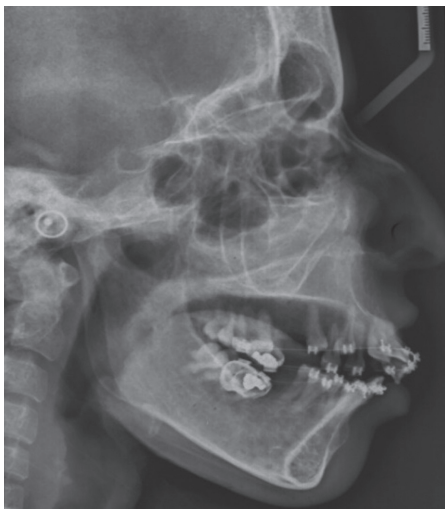

Figure 5. Pre-surgery lateral cephalometric radiograph.

Landmarks	Post-treatment
SNA	82°
SNB	74°
ANB	8°
GoGn/Sn	41°
Y-axis	68°
U1–Sn	72°
IMPA	94°
Interincisal angle	116°
Nasolabial angle	94°
Facial convexity (G'-Sn-Po')	15°

Table 2. Post-treatment cephalometrics of the patient.

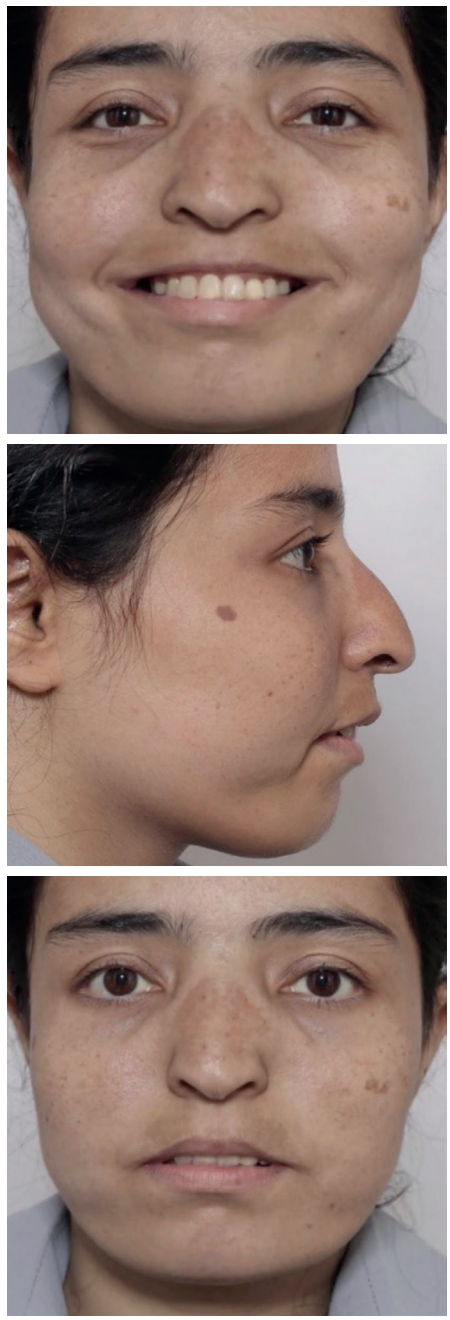

Figure 6. (a–c) Extra-oral post-treatment photographs.

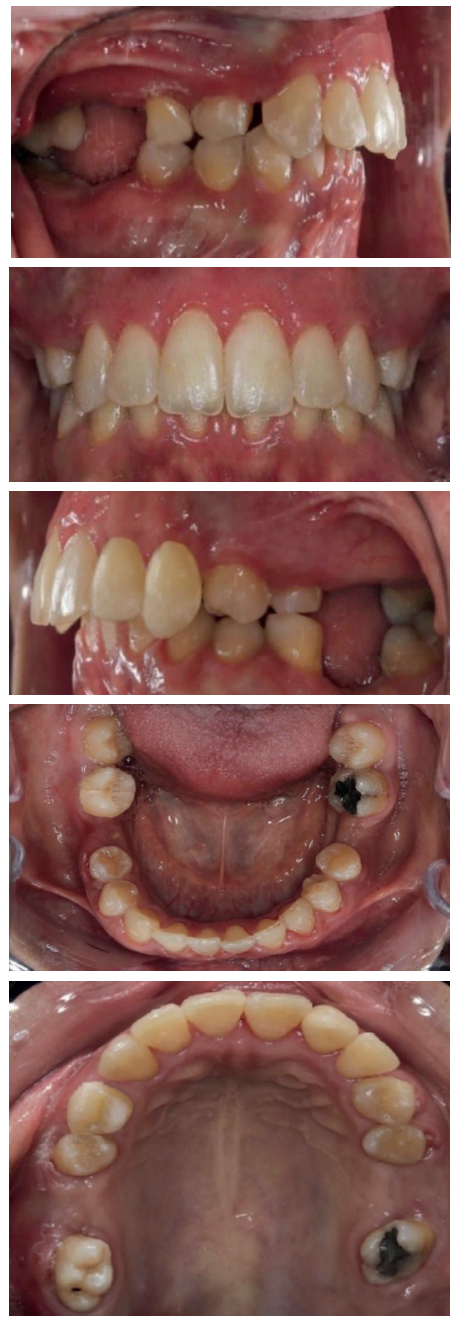

Figure 7. (a–e) Intra-oral post-treatment photographs.

on a comprehensive risk assessment that considered the patient's specific health profile and the surgical team's expertise. Segmental osteotomy, while typically involving less extensive surgical intervention, was deemed to have more risk in terms of potential for significant bleeding given the patient's history of β-thalassaemia major.

The surgical intervention encompassed maxillary impaction and setback, each measuring 6 mm. The removal of the upper third molars was carried out based on the surgeon's judgement. Post-surgery, intermaxillary fixation (IMF) was employed for 6 weeks, followed by an additional 6-week period of intermaxillary elastics to establish normal overjet, overbite, and optimal intercuspation.

The patient refused any periodontal treatment before orthodontics; after the surgical procedure, she was satisfied with her appearance and became highly motivated to pursue treatment. Therefore, she underwent periodontal treatment after the Le Fort 1 osteotomy.

After debonding, she was referred to a prosthodontist for reshaping and composite treatment of the upper and lower teeth, and further treatment. The treatment led to substantial improvements in the alignment of the maxilla relative to the cranial base, effectively addressing the initial concerns of maxillary protrusion and aesthetic imbalance The active treatment phase extended for 4.5 years, culminating in the rectification of maxillary protrusion and a significant enhancement in facial aesthetics (Figures 6 and 7).

The overjet was reduced, although it remained slightly increased, in alignment with the patient's specific craniofacial characteristics that were a result of thalassaemia. The treatment successfully

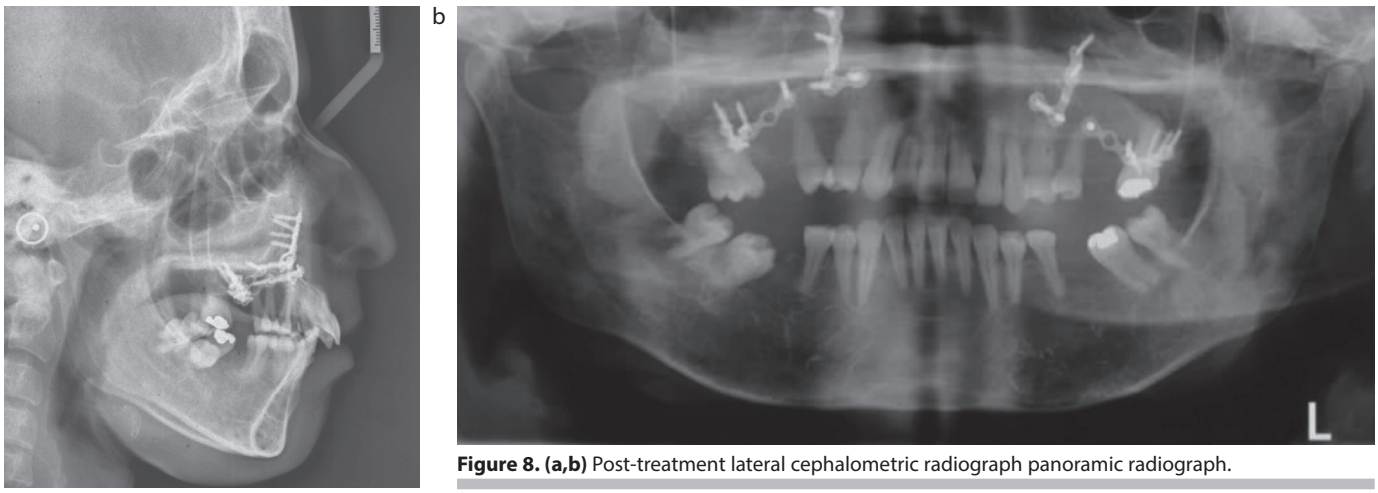

Figure 8. (a,b) Post-treatment lateral cephalometric radiograph panoramic radiograph.

managed the spatial discrepancies initially presented, using orthodontic techniques to align teeth within the new jaw structure. The residual spaces from the extracted teeth were addressed to improve dental function and aesthetics while considering the long-term stability of the results.

These outcomes reflect a considerate adjustment to treatment to manage complexities posed by β-thalassaemia major, emphasizing tailored patient care. Radiographic images are shown in Figure 8. Detailed cephalometric evaluations, encompassing hard and soft tissue analyses, are given in Table 2.

Discussion

β-thalassaemia major is a hereditary disorder marked by profound anaemia, impaired growth, disruptions in endocrine function, and alterations in the skeletal structure resulting from the enlargement and proliferation of haematopoietic marrow, particularly in the maxillary region. Maxillofacial characteristics of this condition include the enlargement of the frontal bones and zygomatic arches in the maxilla, which leads to a concave nasal bridge and outward angulation and spacing on the upper anterior teeth.[8,9,12] The dentofacial irregularity associated with thalassaemia is chiefly related to the maxilla. Consequently, Le Fort osteotomy is the most commonly employed method for addressing this concern. Hypotensive and haemodilution methodologies were employed during the surgical procedure to prevent excessive bleeding and aid in managing blood loss.[4]

In this case, the spaces formed by the absence of teeth were resolved using fixed orthodontic appliances. Alternatively, employing implants to close these spaces was also an option.[12] Extraction of the upper third molars was necessary during the surgery, which could lead to the upward displacement of the lower second molars. Dental implants were considered but ruled out owing to the possible complications from β-thalassaemia major, such as excessive bleeding, delayed healing and compromised bone density, all of which could negatively affect the success of implant placement and post-operative recovery. An alternative solution involves the use of a cantilever bridge using lower second molars as abutment teeth.[13] The surgical and orthodontic approach described here reflects consideration of the challenges posed by β-thalassaemia major, emphasizing the necessity for individualized treatment plans that consider both the systemic health risks and the specific dental anomalies associated with the disorder.

Excessive gingival recession after surgery was hypothesized. Furthermore, the patient's compliance had been notably poor, resulting in frequent missed appointments that considerably extended the duration of her treatment. This prolonged treatment period could be an additional factor contributing to periodontal recession, especially in the upper left canine.[15,16] After the LeFort 1 osteotomy, the periodontal recession led to the patient being referred to a periodontist for treatment, particularly on the upper left canine. Additionally, the patient was referred for restorative dental treatment to reshape the teeth. However, she refused further treatment on the posterior teeth. The patient expressed high satisfaction with the outcomes.

Conclusions

Individuals diagnosed with β-thalassaemia major commonly experience pronounced maxillary protrusion, and rectifying this condition through maxillary setback is fraught owing to underlying systemic problems. In this case report, a patient underwent a successful Le Fort 1 osteotomy procedure, yielding favourable aesthetic outcomes. Given the complexities of β-thalassaemia major, continued monitoring and longitudinal studies are recommended to understand the long-term stability of orthodontic outcomes in such patients, particularly considering the potential for relapse as a result of ongoing bone metabolism issues. Future research could explore innovative materials and less-invasive surgical techniques that may offer reduced risk and enhanced healing properties for this patient demographic.

Compliance with Ethical Standards

Conflict of Interest: The authors declare that they have no conflict of interest.
Informed Consent: Informed consent was obtained from the participant included in the article.

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CPD ANSWERS JULY 2024

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