BULLETIN OF STOMATOLOGY AND MAXILLOFACIAL SURGERY
Volume 20, Issue 1

DOI: 10.58240/1829006X-2024.1-150

CASE REPORT

ORTHODONTIC MANAGEMENT OF THALASSEMIA INTERMEDIA – A CASE REPORT

Prof. Sonali Deshmukh,[1] Dr Asmita Kharche,[1*] Dr. Nandalal Toshniwal[2]

1. Department of Orthodontics & Dentofacial Orthopedics, Dr. D.Y. Patil Dental College and Hospital, Dr. D.Y. Patil Vidyapeeth, Pune 411018, India
2. Rural Dental college of Pravara Institute of Medical Institute of Medical sciences, Loni

** Corresponding author: Dr. Asmita Kharche, Department of Orthodontics & Dentofacial Orthopedics, Dr. D.Y. Patil Dental College and Hospital, Dr. D.Y. Patil Vidyapeeth, Pune 411018, India;*
e-mail: asmita.kharche@dpu.edu.in

Received: *Jan 15, 2024;* ***Accepted:*** *Mar 15, 2024;* ***Published:*** *Mar 25, 2024*

Abstract

Background: Beta Thalasssemia (βT) is a genetic disorder causing various orofacial manifestations. It presents clinically in the form of maxillary prognathism and mandibular retrognathism with steep mandibular plane angles.
Case Description: This case report presents 9.5 yrs male patient with β Thalasssemia and clinically a developing Class II malocclusion with vertical maxillary excess causing a gummy and unpleasant smile. Orthodontic treatment aimed at restricting the growth of maxilla with use of headgear followed by correcting the dental malocclusion with the help of fixed orthodontic. 3M forsus resistance device was given for mandibular advancement and Class II correction
Conclusions: Early intervention, proper diagnosis and ideal treatment strategy of the case helped achieving good occlusion for the patient and controlled the Class II growing tendency typically seen in thalassemia patients if not addressed at an early age.

Keywords: *β Thalassemia; Headgear appliance; 3M Forsus Resistance Device; corrective orthodontics.*

Introduction

Thalassaemia is caused by abnormal hemoglobin, due to gene mutation controlling alpha or beta globins production. Abnormally unpaired globin chains lead to haemolytic anaemia by causing membrane damage and cell death within organ systems and destruction of erythroid precursors in the bone marrow. Thalassemia's origin is dated 50,000 years, in a valley South of Italy and Greece covered by Mediterranean Sea, thus the name thalassemia which has been derived from a Greek word "Thallus" meaning sea. Thomas B. Cooley (1927), was one of the pioneers who reported seven splenomegaly cases with anemia, characteristic bone and facial changes.[1,2]

β-thalassemias are characterized by deficient or absent synthesis of the β-globin subunit of hemoglobin molecule[3] β-thalassemia is inherited as a mendelian recessive gene depending upon which it can be homozygous or heterozygous. Homlogous β-thalassemias presents itself in either a severe anemia form or a mild variant designated as β- thalassemia intermedia.[4] Extraoral appearance of thalasssemia intermedia patients is characterized by pallor, mild jaundice, frontal bossing, orbital hypertelorism and other abnormalities of facies secondary to marrow expansion and abdominal enlargement due to hepatospenomegaly.[5] The orofacial manifestations of thalassemia are mainly due to inefficient erythropoiesis, with the formation excessive

erythroid mass causing bone expansion. Malocclusion seen in thalassemic patients are consequence of severe maxillary protrusion, which results in increased overjet and anterior open bite intraorally and malar prominence, saddle nose, frontal bossing extraorally giving an appearance of chip munk or rodent facies.[6]

There is lack of evidence-based clinical guidelines and research data regarding dental treatment of people with haemoglobinopathies. Thalassemic patients with the need for orthodontic treatment had worse scores for Oral Health related Quality of Life (OHRQoL) compared to the non-affected patients, hence orthodontic treatment should be at priority for improving the oral health and quality of life.[7]

In this present manuscript we intend to show the advantage of starting early orthodontic treatment for controlling the severity of typical characteristics seen in thalassemic patients at later age. this case reports highlights the advantage of using light orthopedic and orthodontic forces for the treatment in contrast to conventional treatment, considering the ill effects of higher forces in thalassemic patients.

Case Presentation

"All procedures performed in this study were in accordance with the ethical standards of the institutional and/or national research committee(s) and with the Helsinki Declaration (as revised in 2013). Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the editorial office of this journal."

A 9.5 year old male visited Smilex Dental Clinic for esthetic and functional correction. Detailed Case history revealed the patient had beta thalassemia intermedia of a mild variant, for which he had received blood transfusions in the early childhood. Chief complaint of the patient was excessive display of upper incisor and a history of trauma to upper left central incisor. On cephalometric evaluation with the use fo Dolphin 2D software Dolphin imaging software® (Dolphin Imaging and Management Solutions, Chatsworth, California, USA) as shown in Table 1 patient presented a comparatively orthognathic maxilla (SNA=78°, A to Nasion perpendicular = -2 mm) as compared to a severely retruded mandible relative to the cranium, (SNB=69°, Pog to Nasion perpendicular = -13mm) causing a skeletal Class II pattern (Wits appraisal of 6.9 mm and ANB - 9°) and hyperdivergent facial growth pattern (FMA = 35°, Go-Gn to SN = 49°, Y Axis to FH = 65°).

Dentoalveolar findings showed Angle's Class II division 1 malocclusion (8) with increased overbite and overjet of 6.0 mm and 9 mm respectively. Skeletal discrepancy showed dental compensation in the form of relatively normal inclination of maxillary incisors (upper incisor to SN =102°, to NA=24° and 4 mm), as well asmandibular incisors proclination (IMPA = 96.2, lower incisor to GoMe =96.2°, lower incisor to NB=35° and 8 mm) (Figure 1A Pre-treatment OPG & Lateral Cephalogram).

Soft tissue finding presented thalassemic facial appearance with a skeletal class II profile, incompetent lips, lower lip trap and a convex facial profile. Complete exposure of maxillary incisors and excessive gingival display was presented in rest position due to short upper lip.

Treatment Objectives

Skeletal
Correction of vertical maxillary excess
Correction of retruded mandible
Class II skeletal discrepancy

Dental
Correction of Class II molars
Spacing and proclination in upper anteriors
Correction of excessive incisor display
Correction of proclined lower anteriors
Ellis Class 2 fracture 21

Soft Tissue
Correction of short upper lip

Treatment Plan
High pull headgear
Extraction of upper and lower first premolars
Fixed functional appliance
Root canal treatment and restoration of non-vital 21
Lip lengthening procedure

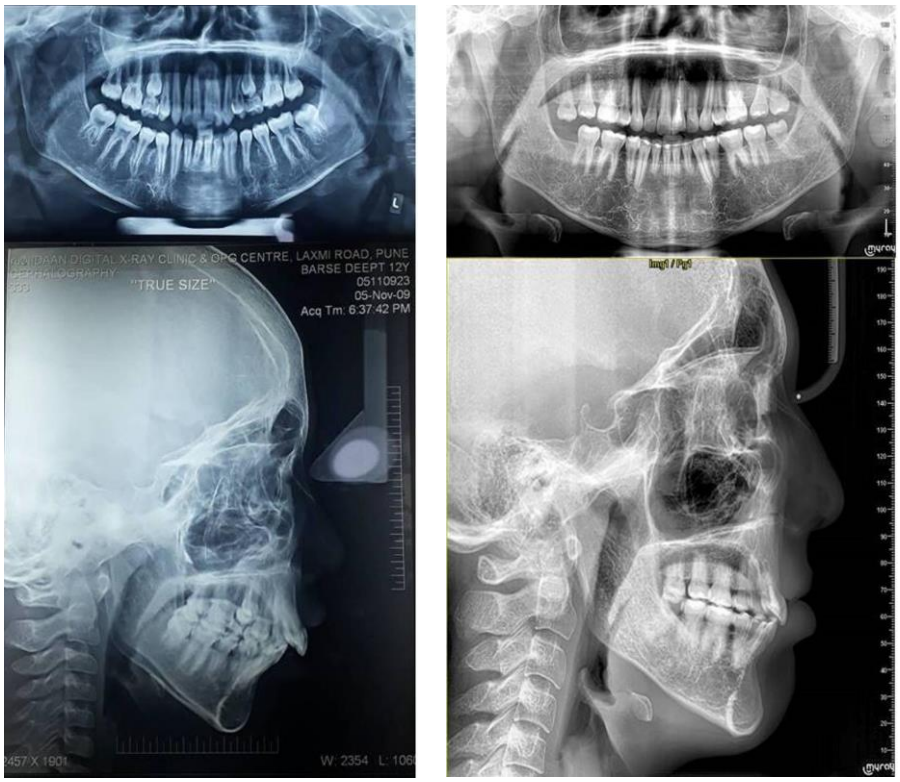

Figure 1. *Pre-treatment* ***(A)*** *and Post-treatment* ***(B)*** *OPG & Lateral Cephalogram*

Orthodontic procedure

As the patient reported during pre-pubertal phase and showed severe skeletal discrepancy it was decided to start growth modulation therapy to improve facial appearance using orthopedic and functional treatment. A high pull headgear with extraoral traction force of 350gm per side was given for the first 5 months, to retract and restrict maxillary growth. The direction of force vector was passed through the center of resistance of maxilla, producing a translatory force to control and improve vertical and sagittal growth of maxillary complex. Patient was instructed to wear headgear for 14-16 hours per day. The lack of patient cooperation and irregular visits made it difficult to track actual wear time and effects of headgear on dentoskeletal effects. Hence after 5 months of orthopedic treatment, fixed orthodontic treatment along with extraction of all 1st premolars was initiated with .018 Roth prescriptions in order to correct the malocclusion and improve overall patient aesthetics [Fig. 2 Intraoral images]. Extraction of all 1st premolars was planned followed by Class II correction using a fixed functional appliance therapy. Forsus fixed functional appliance (3M Unitek) was given for correction of Class II skeletal discrepancy post space closure (Figure 2 Intraoral images of Forsus fixed functional appliance in place).

Appliance was kept in place for 8 months period till overcorrection was achieved in order to compensate for relapse. Total duration of treatment for fixed orthodontics and fixed functional appliance treatment was of 30 months. Retention protocol included maxillary and mandibular fixed retainers and essix removable retainers.

Results

Total orthodontic treatment lasted for 35 months. Facial profile was improved due to restriction of maxillary growth and correction of severely retruded mandible. Fixed orthodontic treatment combined with fixed functional appliance resulted in alignment and leveling of maxillary and mandibular dental arches, correction of class II molar relationship to a stable Class I molar relation, and reduction of the excessive overjet and deep overbite [Figure 2C Post-treatment Intraoral images and Figure 1B Post-treatment OPG & Lateral cephalogram] Table 1 represents pretreatment and post treatment cephalometric measurements. Superimposition of the pre and post treatment cephalometric tracings (Figure 3 Superimposition of pre and post treatment tracing)

showed a maxillary restraint (A to N Perp. Initial -2.0 mm, final -3.5 mm) and an increased mandibular length of 15 mm (CoGn Initial = 94 mm, final = 109 mm) during treatment. Maxillary incisor retraction of 3° and 1.0 mm was achieved along with retraction of mandibular incisor by of 6° and 2.0 mm. The combined effect of maxillary growth restrains and maxillary incisor retraction was reflected in the form of increased nasolabial angle by 4° (initial 95°, final 99°).

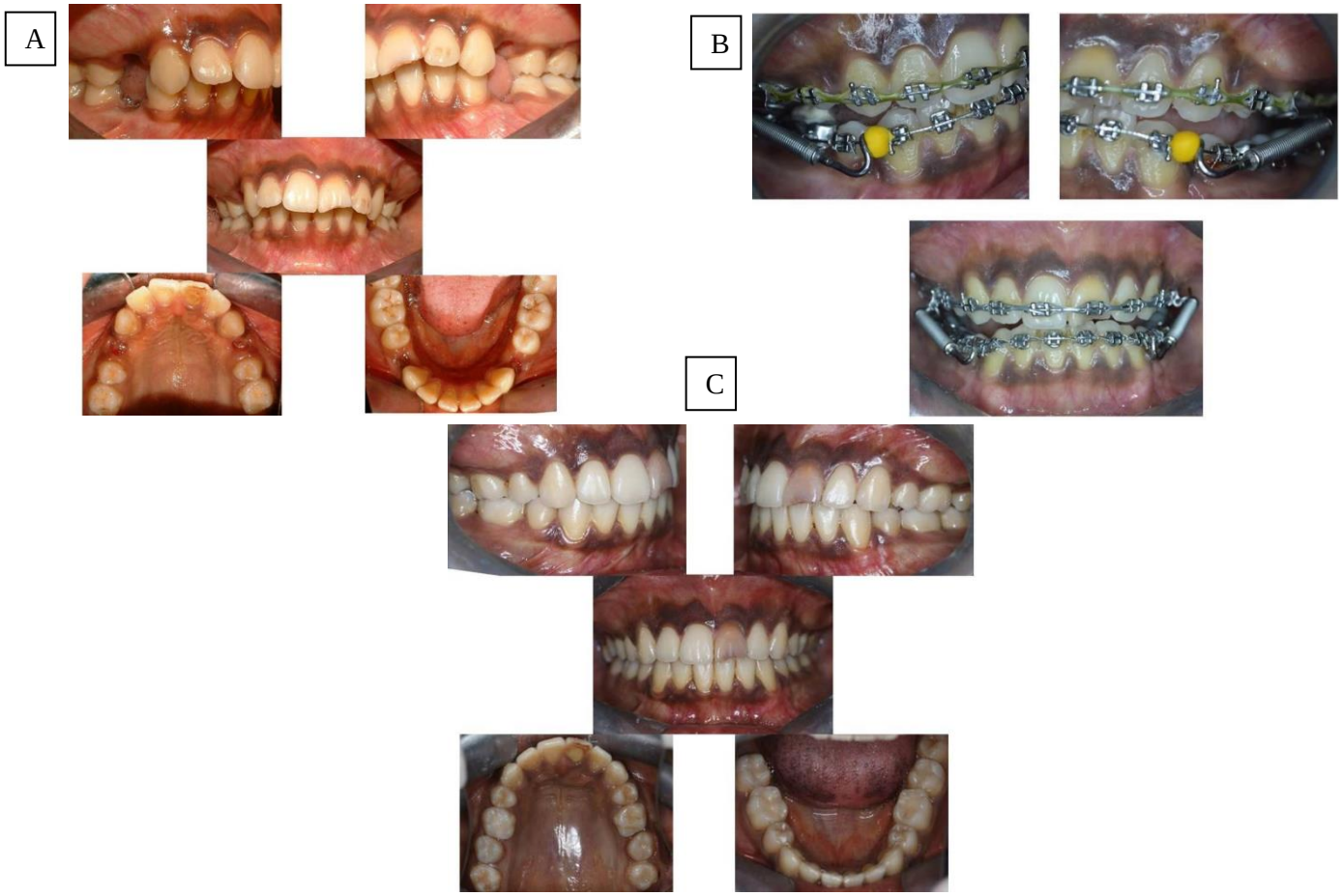

Figure 2. *Intraoral Images After Extraction (**A**), during treatment with Forsus fixed functional appliance in place (**B**) and Post Treatment (**C**)*

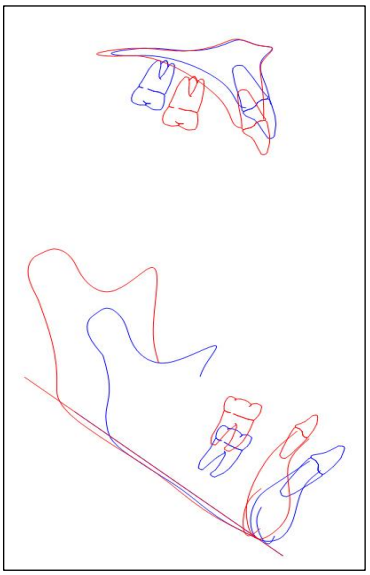

Figure 3. *Superimposition of pre and post treatment tracings (blue – pretreatment tracing and red - post treatment tracing)*

At the end of the treatment moderate resorption was noted at the roots of upper molars and lower incisors due to distally directed forces on upper molars and mesial forces on lower incisors using fixed functional treatment. Extraction was advised for extruded upper right third molar.

Table 1. *The Pretreatment, Mistreatment and Post treatment cephalometric analysis*

Measurement	Norm	Pre	Mid	Post
SNA	82±2	78	77	77
Co-A	84-92mm	76.8	75.22	76.8
A to N perp	0-1mm	-2	-2.9	-3.5
SNB	80±2	69	72	73.1
Co-Gn	98-108mm	94.1	99.19	109
Pog-N perp	-2 to 4mm	-13mm	-8	-4.4
ANB	2	9	4	2.9
WITS	0	6.9	4	2.5
FMA (25°)	25	35	38.5	39
Go – Gn to SN (32°)	32	46	46	46.1
Y axis (FH) (59°)	59.4	64.9	67.3	67.2
PFH/AFH (59-63%)	65	55.7	59.6	56.3
Saddle Angle 132°	123	123	121	121
Articulare Angle 140°	141	156	151	147
Gonial Angle	124	136	140	136
Upper 1 to NA	22/4	24/6	21/4	21/4
Upper 1 to SN (104°)	102	102	96	96
Upper 1 to NF	38.1mm	30.4	36.9	36.7
Upper 6 to NF	26.5	19.5	26.3	26.5
IMPA (90°)	90	96.2	85	91.8
Lower 1 to NB (25°/4)	25/4mm	35/8mm	23/5	29/6
Lower 1 to MP	39.8mm	37.2	39.9	39.8
Lower 6 to MP	33.6	23.8	32.7	33.6
Interincisal	135	112.7	130	125.7
Nasolabial (90°-110°)	90-110	95	99	98.9
Soft Tissue (N -Perp to Pog)	-2mm	-8mm	-5	-3
*E line: U lip	-4mm	2.7	-2	-3.6
*E line: L Lip	-2mm	6.3	1.15	0.74

Discussion

In thalassemic patients, orthodontist must closely monitor tooth movements at shorter intervals, both clinically and radiographically, and use lighter forces than usual as thinning of cortical bone may hasten tooth movement.[9], so the patient had 33 appointments in 27 months of active fixed orthodontic treatment phase.

Thalessemic patients exhibit combination of maxillary hyperplasia and mandibular deformity as described by Bassimitci et al.[10] confirmed the findings of an hyperdivergent facial pattern, steep mandibular planes angle, increased anterior facial height and a reduced total ramal and effective mandibular length leading to a Class II skeletal pattern. Reduced posterior facial height is attributed to anemia which results in growth retardation at the condylar region.[11]

Our patient exhibited increased gingival display due to short lip as well as vertical maxillary excess and also high articular angle (=156°), which affected the vertical dimension and was exhibited by an increase in the vertical cephalometric parameters (Go–Gn to SN=46°, FMA=35°) and reduction in posterior facial height (PFH/AFH=55.77%). The above findings and considering age of the patient, it was decided to start with interceptive treatment by giving a high pull headgear to restrain the maxilla and prevent further vertical growth.

Due to lack of patient cooperation the treatment modality was changed to fixed orthodontic treatment with decision to extract all first premolars in order to correct the soft tissue profile by correcting excessive maxillary anterior display and mandibular incisor proclination. As thalassemic patients exhibit thinning of cortical bone light orthodontic forces were used for retraction and use of temporary anchorage devices were avoided.

Since in thalassemic patients it is recommended to use functional appliances based on muscular origin and minimal effects on anchorage[12] and as our patient showed delayed overall growth spurts fixed functional treatment with 3M™ Forsus™ was then initiated to correct severe mandibular retrusion which lead in correction of maxilla-mandibular ANB discrepancy by 5 degree (ANB – Initial -8 to final – 2.9) and increased in effective mandibular length by 15mm (initial -94mm and Final – 109mm).

In our reported case the combined functional orthopedic not only contributed to restore skeletal harmony and reduced severity of malocclusion, but also to a marked improvement in oral functions and esthetics. Although upper lip remains short and patient was advised lip lengthening procedure after completion of treatment and restoration of upper left central incisor Delaying skeletal treatment and opting for orthognathic surgery is not recommended in thalassemic patients, due to the blood transfusion complications, which is required in these procedures, as well as stability of treatment is questionable due to thin cortical plates.[13] Since the observation is based on a single individual the study limits in generalizing the findings to broader population, as well as patient co-operation fails to provide long term follow up of the case.

Conclusion

Early diagnosis and management are crucial in controlling the severity of craniofacial changes by correcting the affected craniofacial growth. Our βT patient underwent combination of functional orthopedic interceptive treatment which controlled the skeletal changes, followed by an orthodontic treatment and fixed functional stage to achieve proper functional occlusion and improved aesthetics. Such early intervention not only reduces the need for future orthognathic surgery but also contributes in reducing emotional trauma to patient especially in growing years of life by improving facial aesthetics.

Declarations

Conflicts of interest and financial disclosures

All the authors have completed the ICMJE uniform disclosure form (available at https://fomm.amegroups.com/article/view/10.21037/fomm-21-111/coif). The authors have no other conflicts of interest to declare

Ethical approval

"The study was conducted in accordance with the Declaration of Helsinki (as revised in 2013). Patient permission for publication of pictures and data was obtained." "All procedures performed in this study were in accordance with the ethical standards of the institutional and/or national research committee(s) and with the Helsinki Declaration (as revised in 2013). Written informed consent was obtained from the patient for publication of this casereport and accompanying images."

Source of funding

This research received no external funding.

Key findings

Management β Thalassemic Patient showing Class II malocclusion with maxillary prognathism and mandibular retrognathism, by orthopedic and corrective orthodontics.

What is known and what is new?

Report here about what is known.

Report adds to the importance of early intervention by help of orthopedic and corrective orthodontics in improving orofacial manifestations of β Thalassemic, reducing the need of orthognathic surgery.

What is the implication, and what should change now?

Awareness regarding advantage of orthopedic treatment and growth modulation at early age should increase so that patients seek care and limits the need of complex procedures in thalassemic cases.

REFERENCES

1. Cooley TB, Lee P. A series of cases of splenomegaly in children with anemia and peculiar bone changes. *Trans Am PediatrSoc*. 1925;37:29-30
2. Cooley TB, Witwer ER, Lee P. Anemia in children with splenomegaly and peculiar change in bones: report of case. *Am J Dis Child*. 1927;34:347-363. doi:10.1001/archpedi.1927.04130210022002
3. Weatherall DJ. The thalassemias. En: Beutler E, Lichtman MA, Coller BS, Kipps TJ, editors. Williams´hematology. *New York: McGraw-Hill*. 1995:581-615.
4. Cao A, Galanello R 2010. β-Thalassemia. *Genet Med*. 2010;12:61–76. doi:10.1097/GIM.0b013e3181cd68ed
5. Nienhuis AW, Nathan DG. Pathophysiology and Clinical Manifestations of the β-Thalassemias. *Cold Spring HarbPerspect Med*. 2012;2(12):a011726. doi:10.1101/cshperspect.a011726
6. De Mattia D, Pettini PL, Sabato V, Rubini G, Laforgia A, Schettini F. Oromaxillofacial changes in thalassemia major. *Minerva Pediatr*. 1996;48:11-207
7. Tabesh A, Abbasi F, Shavakhi M, Mahmood M. Oral health-related quality of life and orthodontic treatment need in thalassemia major patients. *Dent Res J*. 2022;19:70
8. Angle EH. Classification of malocclusion. *Dent Cosmos*. 1899;41:248-64, 350-7
9. Einy S, Hazan-Molina H, Ben-Barak A, Aizenbud D. Orthodontic consideration in patients with beta-thalassemia major: case report and literature review. *Journal of Clinical Pediatric Dentistry*. 2016;40(3):241-6. doi:10.17796/1053-4628-40.3.241
10. Bassimitci S, Yucel-Eroglu E, Akalar M. Effects of thalassaemia major on components of the craniofacial complex. *Br J Orthod*. 1996;23:157-162. doi:10.1179/bjo.23.2.157
11. Roussos P, Mitsea A, Halazonetis D. et al. Craniofacial shape in patients with beta thalassaemia: a geometric morphometric analysis. Sci Rep. 2021;11(1):1686. doi:10.1038/s41598-020-80234-z
12. Graber T, Vanarsdall R. Orthodontics, Current Principles and Techniques. *The Mosby Co*. 2005;28(2):197. doi:10.1093/ejo/cji127
13. Showkatbakhsh R, Behnia H, Jamilian A, Heydarpour M, Fetrati A. Lefort 1 osteotomy in a beta-thalassemia major patient--a case report. *Int J Orthod Milwaukee*. 2013;24(2):15-18

ԹԱԼԱՍԵՄԻԱՅԻ ԻՆՏԵՐՄԵԴԻԱ ՕՐԹՈԴՈՆՏԻԿ ԿԱՌԱՎԱՐՈՒՄ – ԴԵՊՔԻ ՀԱՇՎԵՏՎՈՒԹՅՈՒՆ

Պրոֆեսոր Սոնալի Դեշմուխ,[1] դոկտոր Ասմիտա Խարչե,[1] դոկտոր Նանդալալ Թոշնիվալ[2]

1. Օրթոդոնտիայի և դիմածնոտային օրթոպեդիայի ամբիոն, Դոկտոր Դ. Յ. Փաթիլ Ատամնաբուժական քոլեջ և հիվանդանոց, դոկտոր Դ. Յ. Փաթիլ Վիդյափեթ, Պունա 411018, Հնդկաստան
2. Պրավարայի բժշկական գիտությունների բժշկական ինստիտուտի ատամնաբուժական քոլեջ, Լոնի, Հնդկաստան

Ամփոփում

Նախապատմություն. Բետա թալասեմիան (βT) գենետիկ խանգարում է, որն առաջացնում է բերանա-

դիմային տարբեր դրսևորումներ: Կլինիկականորեն այն դրսևորվում է ծնոտի պրոգնատիայի և ստորին ծնոտի ռետրոգնատիայի տեսքով՝ ստորին ծնոտի հարթ անկյուններով:

Դեպքի նկարագրությունը. Այս դեպքի զեկույցը ներկայացնում է 9,5 տարեկան արական սեռի պացիենտ β թալասեմիայով և կլինիկական զարգացող II դասի թերակլուզիայով՝ դիմածնոտային ուղղահայաց ավելցուկով, որն առաջացնում է լնդային և տհաճ ժպիտ: Կատարվել է օրթոդոնտիկ բուժում, որի նպատակն էր սահմանափակել վերին ծնոտի աճը, որին հաջորդել է կցվածքի շտկումը ֆիքսված օրթոդոնտիկ ապարատի օգնությամբ: 3M forsus դիմադրության սարքը տրվել է ստորին ծնոտի առաջխաղացման և II դասի ուղղման համար

Եզրակացություններ. Վաղ միջամտությունը, ճիշտ ախտորոշումը և դեպքի բուժման իդեալական ռազմավարությունը օգնեցին հիվանդի համար լավ կցվածքի ստացմանը և վերահսկեցին II դասի աճի միտումը, որը սովորաբար նկատվում է թալասեմիայով հիվանդների մոտ, եթե վաղ տարիքում չլուծվի:

ОРТОДОНТИЧЕСКОЕ ЛЕЧЕНИЕ ПРОМЕЖУТОЧНОЙ ТАЛАССЕМИИ – ОТЧЕТ О СЛУЧАЕ

Профессор Сонали Дешмукх,[1] доктор Асмита Кхарче,[1] доктор Нандалал Тошнивал[2]

1. Кафедра ортодонтии и зубочелюстной ортопедии, Доктор Д.Ю. Патил Стоматологический колледж и больница, доктор Д. Ю. Патил Видьяпит, Пуна, 411018, Индия
2. Сельский стоматологический колледж Медицинского института Правара, Лони,Индия.

Абстракт

Введение: Бета-талассемия (βT) — генетическое заболевание, вызывающее различные орофациальные проявления. Клинически он проявляется в виде прогнатизма верхней челюсти и ретрогнатизма нижней челюсти с крутыми углами плоскости нижней челюсти.

Описание случая: В этом отчете о болезни представлен 9,5-летний пациент мужского пола с β-талассемией и клинически развивающейся аномалией прикуса II класса с вертикальным избытком верхней челюсти, вызывающим липкую и неприятную улыбку. Ортодонтическое лечение, направленное на ограничение роста верхней челюсти с использованием головного убора с последующей коррекцией неправильного прикуса с помощью несъемных ортодонтических аппаратов. Устройство сопротивления 3M forsus было предоставлено для продвижения нижней челюсти и коррекции класса II.

Выводы: Раннее вмешательство, правильный диагноз и идеальная стратегия лечения данного случая помогли добиться хорошей окклюзии у пациента и контролировать тенденцию к росту II класса, которая обычно наблюдается у пациентов с талассемией, если не лечить ее в раннем возрасте.