



Soft Tissue Profile Analysis for Iraqi Patients with β -Thalassemia Major

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Abstract

The facial appearance of patients with β -thalassemia major is readily recognizable so that soft tissue profile analysis is presented in this study.

The aim of the study is concerned about the analysis of soft tissue profile morphology in a group of patients having β -thalassemia major and to record the possible differences between both thalassemic genders and to investigate the differences in the soft tissue morphology between thalassemic patients and non thalassemic normal subjects

The material of this study consisted from lateral cephalometric radiographs for (40) thalassemic patients aged 15-17 years and (40) cephalometric radiographs for non thalassemic normal subjects were obtained for analyzing soft tissue profile morphology

The results showed that there were no significant differences between thalassemic males and females but the soft tissue profile morphology for thalassemic patients mostly different from normal subjects which characterized by convex profile.

Thalassemic patients have a soft tissue profile morphology mostly different from normal's that characterized by smaller nasal dimensions, short and everted upper lip and thicker lower lip which giving them the appearance of convex profile.

Key words: β -thalassemia major-soft tissue profile-cephalometric.

Introduction

Thalassemia can be defined as a heterogeneous group of inherited disorder characterized by reduced or absent synthesis of one or more types of normal hemoglobin polypeptide chain in another ward these hemolytic disorders are caused by a partial or complete deficiency of α or β globin chain synthesis ¹.

It is now clear that thalassemia probably represents the most common genetic disorder causing a major public health problem in the world's population, thalassemia has important

orthodontic implications, which is rarely seen in the orthodontic practice, and consequently, little attention is paid to it. Nevertheless, it is important to record whatever experience is accumulated regarding orthodontic diagnosis and treatment for such condition², as a result, it could be suggested that the skeletofacial morphology of thalassemic patients could be different from the general population and that was proved by many studies^{3,4,5}.

The course of the disease is depends on the child is maintained on an adequate transfusion program and in

the absence of transfusion therapy the clinical course is characterized by severe anemia ,retardation in the rate of growth and development, body disproportion with truncal shortening, hepatosplenomegaly and cortical thinning lead to pathological fractures^{6,7}.

The bimaxillary expansion one of the classical clinical changes of the disease, the progressive maxillary enlargement, maxillary dysplasia, separation of the orbit, upward protrusion of the lip and exposing upper teeth, severe protrusion of the middle third of the face, prominence of the zygomatic bones tend to obscure the base of the nose and frontal bossing all these disturbances of the craniofacial growth give rise to the classical "chipmunk" faces^{4,8,9}.

There were increase in the mandibular vertical height and a tendency to disto-occlusion were found between children with thalassemia major, expressed by large overjet result from changes in arch width and anterior movement of the maxilla^{9,20}.

Comprehensive evaluation of a patient's malocclusion and facial balance would be incomplete without inclusion of the soft tissue component, and analysis of both dental and skeletal patterns alone may provide inadequate or misleading, for marked variation exists in the soft tissue covering the skeletodental framework.⁽¹⁰⁾

Traditionally, it has been assumed that facial contours are primarily the result of underlying hard tissue positioning and the subsequent of tissue drap^{11,12}.

While it has been shown previously that the soft tissue vary in thickness over different parts of the facial skeleton, consequently, the outline of the soft tissue profile dose not correspond well with the underlying skeletal framework^{10,13} so that complete analysis of facial profile,

therefore, should also include an evaluation of soft tissue morphology

Materials and method

The sample used in this work consisted of 40 patients with β -thalassemia major (20 males ,20 females) aged 15-17 years which selected from a larger group of patients who attended to thalassemia center in Al- Karama hospital and Ibn- Albalady hospital as they fulfill the criteria of clinical sample selection that includes

- 1-They are patients with β -thalassemia major proved by laboratory and medical examination
- 2-Iraqi in origin aged 15-17 years
- 3-No present or history of orthodontic treatment and maxillofacial surgery

The other group, control, consisted of 20 patients, their selection based on the following criteria's:

- 1.The sample is selected on the bases of satisfactory facial esthetic matching with age.
2. There is no marked facial asymmetry.
3. There is no any systemic abnormality.
4. Full complement of permanent teeth except third molar.
5. There is no history of orthodontic treatment.

Method

All the subjects examined clinically, then lateral cephalometric radiographs were made using Planmeca machine, with the patient in rest position (minimal lip activity and teeth in contact). The film was placed on viewer with the image directed to the right, then tracing following the out line of the soft tissue profile contour from the upper part of the head to the lower part of the chin with the external and internal contour of the cranium, orbit , maxilla, mandible were drawn

on tracing film ⁽¹²⁾ reference landmarks were digitized for subsequent measurements of (12) linear and (2) angular dimensions, these landmarks were located according to the definitions of Brustone⁽¹⁰⁾, Nanda et al¹⁴, Riolo et al¹⁵, and Bishara¹⁷. **Fig.(1)**

Cephalometric landmarks

1. **Pronasale (Prn)**: the most anterior point of the nose in the median sagittal plane.
2. **labrale superius (Ls)**: the point at the superior margin of the upper membranous lip in the median sagittal plane.
3. **labrale inferius (Li)**: the point at the superior margin of the lower membranous lip in the median sagittal plane.
4. **Subnasale (Sn)**: the point at which the nasal septum merges mesially with the integument of the upper lip.
5. **Subspinale (point A)**: the deepest midline point in the curved bony outline from the base to the alveolar process of the maxilla.
6. **Supramentale (point B)**: most anterior part of the mandibular base, between infradentale and pogonion.
7. **Stomion (sto)**: central point of the interlabial gap.
8. **Skin N`**: is located at the point of the maximum convexity between nose and forehead.
9. **Soft tissue pogonion (pog`)**: the most prominent or anterior point on the chin in the mid sagittal plane.

Frankfort horizontal reference plane and vertical plane on the Frankfort horizontal plane from the posterior border of the pterygomaxillary fissure are used as a vertical and horizontal reference planes.

Statistical analysis

The data of the sample was projected to statistical analysis using Spss version 12 programe

Mean values, standard deviation (SD), P value, and student paired "t test" were calculated

All P values above 5% are regarded a statistically insignificant as followed

$p > 0.05$ NS non significant

$p < 0.05$ S significant

$p < 0.01$ HS highly significant

Results

General look for Table 1 demonstrates that means and p value for thalassemic males and females patients are not statistically significant for all linear and angular measurements.

While Table 2 demonstrated that thalassemic females have a smaller mean value with a higher significant differences for nose depth, nose length, nose height, and both upper and lower lip length than normal females, the lower lip thickness at labrale inferius and point B indicate that thalassemic females have a higher mean value with a highly significant differences.

Table 3 summarizes the comparison between thalassemic males and normal males, thalassemic males have a notable decrease with highly significant differences for nose height, nose length, nose depth, upper lip thickness at point A, length of the upper, lower lip and soft tissue thickness at Pog` and the position of the upper lip to E-line.

The lower lip thickness at labrale inferius and point B indicate that thalassemic males have a significantly higher mean value.

Discussion

The assessment of the balance and harmony of the face includes the morphologic relationships of the nose,

lips, and chin as well as the underlying skeletal components so that the radiographic changes in the skeleto-dental complex and soft tissue profile morphology represents the most striking and consistent findings in this type of hemolytic anemias.

There were many limiting factors affect on the size of the sample selection and the most important one that most of thalassemic patients die in a younger age by intractable heart failure, besides that, many of patients refuse doing the radiographs due to severe emotional trauma that they sustain during childhood because of their appearance and the radiation may have adverse effects on them.

There was a great deal of variability between both thalassemic females and males and normals (table 2, table 3) particularly in the nose height, nose depth and nose height which were smaller with a significant differences, these findings could be explained according to the findings that the major effects of thalassemia on the facial skeleton results in overgrowth of the bone marrow that impedes the pneumatization of the paranasal bones and causes enlargements of the malar bone, swelling of the zygoma, sunken bridge of the nose and depression of the root of the nose^{4,8}.

The thickness of the upper lip at labrale superius was thinner in both thalassemic females and males than normals without a significant difference while the thickness of the upper lip at point A is thinner in both thalassemic genders than normals (table 2, table 3) the thickness of the upper lip was thinner in class II due to angulation of the incisors Brustone¹⁰, thalassemic patients have class II malocclusion^{3,5} so this finding explain why thalassemic patients possess a thinner upper lip than normals.

Both thalassemic males and females possess a thicker red part of the lower lip at labrale inferius and increased soft tissue thickness at point B with a significant differences, the thickness measurements is greatly affected by the type of skeletal factor, it seems to be increased in cases of class II due to the increased overjet which will put the upper part of the lower lip away from the labial aspect of the lower incisors, Brustone¹⁰, stated that the increased value of labrale inferius can be partly attributed to deflection of the lower lip produced by the overjet, Rakosi¹⁶ reported that the thickness of the lower lip at labrale inferiors increases with class II cases, the above mentioned findings could be explained why thalassemic patients have thicker lower lip, in addition to that, the thickness of the lower lip depends on the position of the mandible and the increase in the gonial angle which indicates more of a tendency to posterior rotation of the mandible and pronounced vertical growth pattern that seemed to be a consistently observable pattern in thalassemic patients^{3,5}, and could be considered the responsible cause for increased thickness of the lower lip.

The thalassemic females and males data reveals more decreased of thickness at the soft tissue pog` as viewed from the observations for both genders, the reduction in thickness at the soft tissue chin may have been attributed due to the increase in the gonial angle with the vertical growth pattern^{3,5,18} causing slight stretching of the soft tissue over the chin.

Upper lip length was shorter with a significant difference for both thalassemic females and males (table 2, table 3), class II skeletal pattern¹⁷ and vertical growth tendency with the protrusion of the upper incisors⁴ and short upper lip all these factors have an influence on the upper lip to be

retracted and everted, this findings correspond with the findings of ^{18,19}.

Similar explanation could be applied to decreased length of the lower lip in thalassemic females and males due to vertical growth pattern

The position of the upper lip relative to E-line show a significant difference in thalassemic males (table 3) this may be explained according to that the upper lip is thinner and short due to that it seemed to be retracted and slightly everted so it will show recession from the E-line.

It can be summarized the soft tissue profile morphology for thalassemic females and males according to the decrease in nose dimensions together with changes in the chin resulted in noticeable retraction in the lips and retrusive profile that give rise to the appearance of "rodent" faces^{4,18}.

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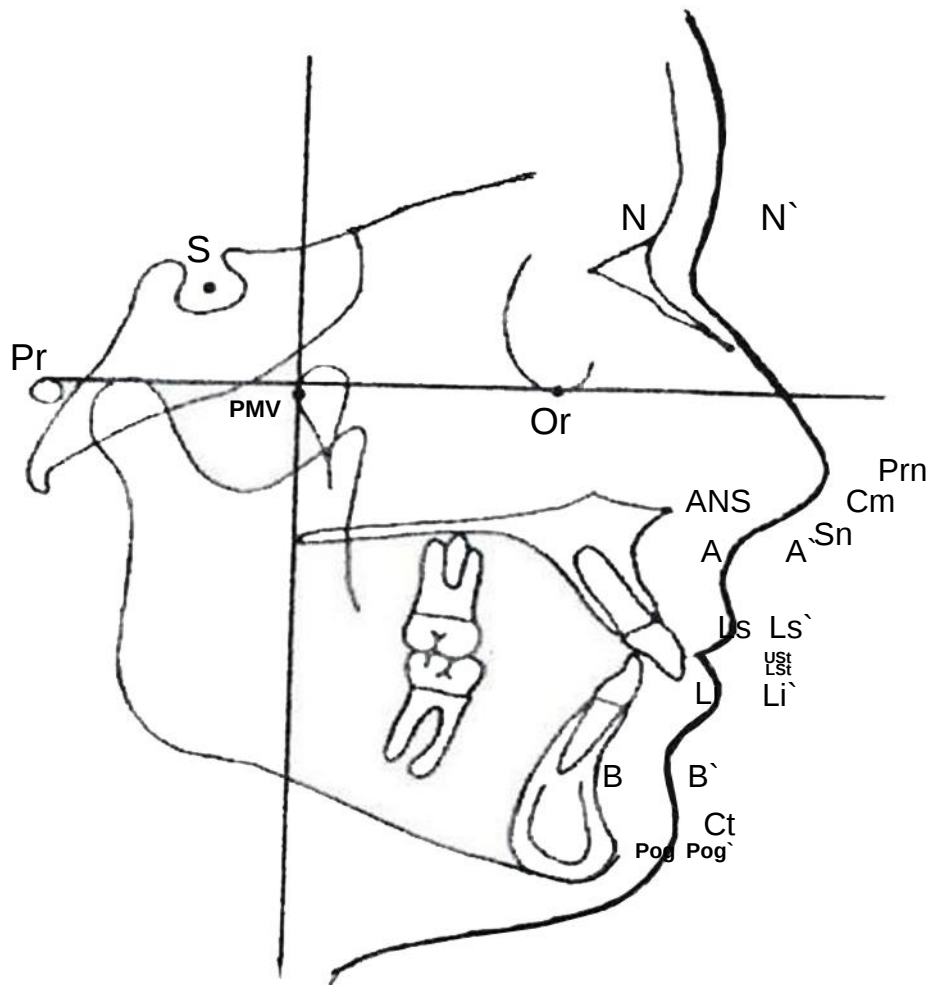


Table 1 soft tissue descriptive statistics for thalassemic males and females

Variables	Thalassemic				t-test	p-value	Sig.
	Males		Females				
	mean	Std. Deviation	mean	Std. Deviation			
Nose height (N'-PMVL thru ANS)	48.8820	2.83011	49.3615	2.89238	-0.530	0.599	NS
Nose length (N'-Prn)	39.8675	2.39673	40.2765	2.40730	-0.538	0.593	NS
Nose depth (PMV-Prn)	78.2265	2.07665	78.0800	2.07665	0.230	0.820	NS
Upper lip thickness at point A (A-A')	12.9240	1.08454	13.0500	1.52881	-0.301	0.765	NS
Upper lip thickness at labarle superius (Ls-Ls')	14.5030	3.02373	12.8105	1.03223	2.369	0.098	NS
Lower lip thickness at labarle inferius (Li-Li')	17.6815	2.59231	18.5970	1.30910	-1.410	0.167	NS
Lower lip thickness at point B (B-B')	12.5945	2.02675	12.2735	0.76818	0.662	0.512	NS
Soft tissue thickness at pogonion (Pg-Pg')	10.2630	1.01086	10.2640	1.05118	-0.003	0.998	NS
Upper lip length (As-PMVL thru Ust)	13.9440	1.97281	13.7980	1.81492	0.244	0.809	NS
Lower lip length (Bs-PMVL thru Lst)	15.4870	2.89983	15.2535	2.93744	0.253	0.802	NS
Upper lip to esthetic planc (Ls-E plane)	2.7285	1.60086	2.9395	1.42336	-1.441	0.662	NS
Lower lip at esthctic plane (Li-E plane)	3.5320	1.39513	3.6645	1.56316	-0.283	0.779	NS
Naso labial angle (Cm-sn-Ls)	111.3200	6.65998	113.3800	8.51553	-0.852	0.399	NS
Mento labial angle (Li-Bs-Ct)	124.6950	7.02753	124.6950	7.11806	-0.300	0.766	NS

Table2 Soft tissue discriptive statistics for thalassemic females and normal females

Variables	Females				t-test	P-value	Sig.
	Thalassemic		Normal				
	mean	Std. Deviation	mean	Std. Deviation			
Nose height (N'-PMVL thru ANS)	49.3615	2.89238	52.0570	3.56149	-0.627	0.012	HS
Nose length (N'-Prn)	40.2765	2.40730	49.5765	1.45539	-14.785	0.000	HS
Nose depth (PMV-Prn)	78.0800	1.95768	79.6120	1.28610	-2.925	0.006	HS
Upper lip thickness at point A (A-A')	13.0500	1.52881	14.1150	0.61619	-2.890	0.006	HS
Upper lip thickness at labarle superius (Ls-Ls')	12.8105	1.03223	12.8835	0.54984	-0.279	0.782	NS
Lower lip thickness at labarle inferius (Li-Li')	18.5970	1.30910	14.1020	0.63594	13.812	0.000	HS
Lower lip thickness at point B (B-B')	12.2735	0.76818	11.0465	0.64068	5.486	0.000	HS
Soft tissue thickness at pogonion (Pg-Pg')	10.2640	1.05118	13.0640	0.39392	-11.155	0.000	HS
Upper lip length (As-PMVL thru Ust)	13.7980	1.81492	16.2180	0.77809	-5.481	0.000	HS
Lower lip length (Bs-PMVL thru Lst)	15.2535	2.93744	17.1635	1.38635	-2.630	0.012	S
Upper lip to esthetic planc (Ls-E plane)	2.9395	1.42336	3.5640	0.77894	-1.721	0.093	NS
Lower lip at esthctic plane (Li-E plane)	3.6645	1.56316	2.8175	0.58482	2.270	0.029	NS
Naso labial angle (Cm-sn-Ls)	113.3800	8.51553	110.3800	4.92508	1.364	0.181	NS
Mento labial angle (Li-Bs-Ct)	124.6950	7.11806	127.1900	4.48587	-1.326	0.193	NS

Table3 Soft tissue discirptive statistics for thalassemic males and normal males

Variables	Males				t-test	p-value	Sig.
	Thalassemic		Normal				
	mean	Std. Deviation	mean	Std. Deviation			
Nose height (N'-PMVL thru ANS)	48.8820	2.83011	52.3490	2.52139	-4.091	0.000	HS
Nose length (N'-Prn)	39.8675	2.39673	50.2690	1.04111	-17.802	0.000	HS
Nose depth (PMV-Prn)	78.2265	2.07665	82.4815	2.14454	-6.374	0.000	HS
Upper lip thickness at point A ') (A-A	12.9240	1.08454	13.5390	0.83934	-2.006	0.052	S
Upper lip thickness at labarle ') superius (Ls-Ls	14.5030	3.02373	13.1955	0.55902	1.902	0.065	NS
Lower lip thickness at labarle ') inferius (Li-Li	17.6815	2.59231	121525	1.57760	8.148	0.000	HS
Lower lip thickness at point B ') (B-B	12.5954	2.02675	11.7955	3.08572	-0.243	0.014	S
Soft tissue thickness at ') pogonion (Pg-Pg	10.2630	1.01086	11.2170	1.11620	-2.833	0.007	HS
Upper lip length (As-PMVL) thru Ust	13.9440	1.97281	16.6080	0.67292	-5.716	0.000	HS
Lower lip length (Bs-PMVL) thru Lst	15.4870	2.89983	17.5500	0.62900	-3.109	0.004	HS
Upper lip to esthetic planc (Ls-) E plane	2.7285	1.60087	3.8390	1.26772	-2.432	0.020	S
Lower lip at esthctic plane (Li-) E plane	3.5320	1.39513	2.8735	1.14636	1.631	0.111	NS
Naso labial angle (Cm-sn-Ls)	111.3200	6.65998	112.0500	6.12969	-0.361	0.720	NS
Mento labial angle (Li-Bs-Ct)	124.0250	7.02753	122.6550	6.43154	0.643	0.524	NS