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Investigating the relationship between major thalassemia diseases with anthropometric sizes of head and facial soft tissue

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ABSTRACT

Literature review and aim: according to increasing prevalence of beta thalassemia (thalassemia major) and based on raised concerns about abnormal head and facial anthropometric besides recognized side effects and etiological importance of these disorders in these sizes and considering that previous studies lacked investigations about relationship between anthropometric sizes of head and facial heard tissues with major thalassemia we decided to perform a research aiming at analysis of the relationship between anthropometric sizes of head and facial soft tissue with major thalassemia and comparing it with normal people.This research was performed by case control method. Case group consisted 30 patients with thalassemia and control group also included 30 normal people who were approximately similar to case group considering their age, gender and BMI. Age range of both groups was from 17-35. Clinical examination, interview, and measuring indicators in both patient and normal group before receiving any treatments including orthodonsy, trauma, plastic operation in head and facial area. Anatomic points were marked with black oily pencil on meant areas of head and facial. Angle sizes were recorded by 0/5-degree approximation and linear sizes were recorded by 0/5 approximation. In this research, anthropometric standard tools for measuring head and face were sliding caliper, measuring tape, Spreading caliper. For comparing both groups, statistical ANOVA test was used and in order to determine statistical difference between groups, t test was used.Statistical results indicated that anthropometric indicators of head and facial soft tissue are affected considering comparison of thalassemia and normal groups. In 20 indicators no significant difference was observed P<0/02 and in 11 indicators reduction happened (p<0.05) and in the next 10 indicators an increase occurred (p<0.05).Due to anemia, patients with thalassemia are suffered from bone marrow hyperplasia in wide bones of head and face such as maxilla, frontal, mandible ramus and other deformations, so transverse growth is happening in bones of these people and consequently their covering soft tissue is growing transversely.

KEY WORDS: MAJOR THALASSEMIA, ANTHROPOMETRIC SIZES, HYPERPLASIA, HEAD AND FACIAL SOFT TISSUE, BONE MARROW

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INTRODUCTION

First time, in 1925, Thomas Cooley who was children professor in Detroit, described this disease. Major thalassemia diseases is an inherited blood disease which is identified by producing abnormal hemoglobin products. Major thalassemia in many countries is being considered as serious risk for public health. Diagnosis and explanation of oral, jaw and facial abnormalities in these patients are dentists and oral disease professional's tasks. Unfortunately, there are few studies on effect of thalassemia on anthropometric sizes. Clearly, in order to create more accurate 3 dimensional descriptions from anthropometric sizes abnormalities in thalassemia patients and to provide a general treatment there is needed to perform many researches (Bassimitici 1996). Since bone marrow is main and essential center for body hematopoiesis, thus during anemia, this center is activated and make up anemia by its activation. So due to increase of bone marrow activation slowly this hematopoiesis center is developed and its size is increased. Since wide head and facial bones such as maxilla all have the increase of activity in major thalassemia patients so, naturally, deformation of bones in this area of head and face cause abnormalities in hard tissue and consequently soft tissue (Karakas 2016).

One side effect of major thalassemia is increase of upper jaw bone growth and also decrease of lower jaw bone growth which finally leads to class two malocculosion (Pakshir, Abu Alhaija 2002, Einy 2016).Thalassemia syndromes are distinct group of hereditary hematopoiesis which their sign was disorder in globin chain production (Amini et al, 2016).

A few studies have been performed, or at least reported, on effect of thalassemia on three dimensional growth of head and face so far. There are also different reports on effect of thalassemia on hard tissue profile which can cause changes in soft tissue. Some researchers reported enlargement of upper jaw is due to overexpansion of red bone marrow and increased bone volume which happen for making up healthy red globules reduction. In contrast, some studies reported that this disease reduces lower jaw growth. However, considering different effects of thalassemia on upper jaw and lower jaw and according to the relationship between changes of hard tissue and soft tissue, this disease makes changes in soft tissue profile.

Based on popularity and increasing prevalence of beta thalassemia, considering concerns for abnormal anthropometric form of head and face and also for identified side effects and ethology of these disorders in these sizes, and based on previous researches we performed a study relationship between anthropometric sizes of head and facial hard tissue with beta thalassemia and compare differences of normal people in Iran professional infirmary of adults thalassemia who were under treatment and control group.

MATERIAL AND METHODS

Population under study included patients aged 17-35 with major thalassemia who were referred to the Iran adults professional infirmary and they were under treatment. As there were some restrictions and some patients did not participate, this research was performed by case control method. Both group are similar considering age, gender based on BMI entry standards. They were attended to the examination after providing information and taking their written consent. Case group included 30 patients with tallassemia (14 females and 16 males with average age of 29/7±4/2 and 30 normal people (15 females and 15 males with average age of 23/7±3/4).

41 linear and angle indicator in facial area were measured by Farkas method. Regarded landmarks were marked with black oily pencil in order not to error happens in measurement. Applied landmarks in this study were standard indicators which are used in standard study of head and facial anthropometric.

In this paper, accurate anthropometric standard tools made in Insize company from Austria were used for measurements which has a metric strip, digital sliding colpier with long jaws, digital coulisse, Digital hook depth gauge, Digital protractor and level. Tools were calibrated and their accuracy was 0.01 mm and 0.01 degree. In general comparison of both groups, in order to determine statistical difference, t test was applied (figure 1).

RESULTS AND DISCUSSION

The research was performed on 30 patients with major thalassemia and 30 people in control group. Under study population aged 17 to 35. Results are shown as below.

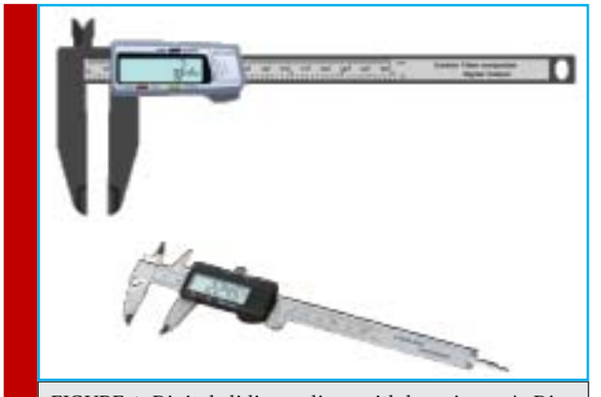

FIGURE 1. Digital sliding coliper with long jaws: A Digital sliding coliper : B

Table 1. 41 linear and angular indices in face area

Measurement area	Index	Millimeter/ degree
Nose width	al-al	
Nose outgrowing rate	sn-prn	
Nose bridge inclination	Nasal bridge (degree)	
Nasolabial angle	Nasolabial angle (degree)	
Mouth width	ch-ch	
Upper lip height	sn-sto	
Height of cutaneous upper lip	sn-ls	
Height of upper lip Vermillion	ls-sto	
upper lip inclination	sn-ls (degree)	
lower lip height	sto-sl	
Height of cutaneous lower lip	li-sl	
Height of lower lip Vermillion	sto-li	
lower lip inclination	sl-li (degree)	
Height of upper part of chin	sl-pg	
Height of lower part of chin	pg-gn	
Chin height	sl-gn	
Throath length	gn-c	
Angle of chin/neck	Mentocer vical angle (degree)	
Measured area	Index	Millimeter/degree
Head round	g-op-g	
Head width	eu-eu	
Head length	g-op	
Head height	v-n	
Forehead height	tr-n	
Height of 1/3 of middle face (nose)	n-sn	Nose height
morphologic height of face	n-gn	
physiognomic height of face	tr-gn	
morphologic height of upper face	n-sto	
Height of 1/3 of upper face (nose)	tr-g	
Height of 1/3 of middle face (nose)	g-sn	Nose height
Height of lower 1third of face	sn-gn	
Mandible height	sto-gn	Height of lower part of face
Face width	zy-zy	
Mandible width	go-go	
Maxilla depth	t-sn	
Mandible depth	t-gn	
Upper face profile inclination	g-sn (degree)	
lower face profile inclination	sn-pg (degree)	
Ear width	pra-pa	
Ear height	sa-sba	
inner corner of the eye space	en-en	
outer the eye space	ex-ex	
Eye length (left)	en-ex	

In order to determine reliability of measurement method, all 41 linear and angular indices were measured for 10 women and 1- mean twice in order not to forget previous recorder numbers by project executive with time intervals. Then correlation between three recorder numbers were studied by intra class correlation. Finally, collected data were analyzed in SPSS software for each person. Shapiro- wilk test cleared that results have normal distribution. Then (mean±SD) was determined for each indicator. After that, Confidence interval of each indicator was estimated in Iranian community by 95% confidence.

As Pvalue higher than 0/05 indicates that the difference is not considerable and it is not identifiable by eyes so it is no clinically significant. Therefore, in this study $p<0.05$ is significant. Considering that normality limit of each indicator is considered as mean±2SD so this was considered as normal rate for Iranian people and it was identified that what percent of the population are out of Iranian adult population.

Case group included 30 patients with thalassemia (14 females and 16 males with average age of 29/7±4/2 and 30 normal people (15 females and 15 males with average age of 23/7±3/4). Both group are similar considering age, gender, trauma record- head and facial operation- orthodonsy treatment, and BMI entry standards and their difference was not significant $p<0.2$.

Table 2. characteristics of people under study in terms of major thalassemia and control groups

BMI		Another disease other that thalassemia which cause change of soft and hard tissues		orthodonsy treatment		Manipulation by head and face surgery		Trauma		gender		Age	Intervening variable / Major thalassemia
		suffered	normal	suffered	normal	suffered	normal	suffered	normal	male	Female		
22/56±3/4		0	30 100%	0	30 100%	0	30 100%	0	30 100%	15 50%	15 50%	23/7±3/4	Normal N=30
20/12±2/1		0	30 100%	0	30 100%	0	30 100%	0	30 100%	16 53/3%	14 46/6%	29/7±4/2	suffered N=30

Head and facial anthropometric indicators of head and neck anatomic points of under study group are provided at the following table and they show that:

1. Out of measured indicators in head area which are provided in table 3, head height index (v-n) showed maximum difference in compare to thalassemia group (thalassemia: 96/99±7/28 and control group: 5/7±105/3 and head width index (eu-eu) represented minimum changes. In addition, out of these indices, head width ($p<0.4$) was not significant statistically.
2. Measured indicators in ear area indicated that out of ear indices, there is no significant difference in earlobe width (par-pa) (thalassemia group: 35/152/7 and control group 35/7±2/6 and earlobe length (sa-sba) (thalassemia group: 59/95±3/136 and control group 61/4±4/1.
3. Out of eye indices, other than both inner corner of the eye (en-en) which do not show significant difference with control group. Other parameters including space between both outer corners of the eye (ex-ex) and eye gap length (en-ex) showed significant difference and space of both outer corners of the eye indicated maximum difference (thalassemia group: 97/29±2/65 and control group 93/5±2/75) in addition, eye gap length of patients with thalassemia (34/72± 2/04) was more that control group (31/39±1/8).
4. From anthropometric indices of nose, nose tip photogene (sn-prn) represented maximum difference which is significant statistically (thalassemia group: 11/80±1/58 and control group 19/6±2/4). While nose bridge inclination showed minimum changes which is not significant statistically. In nose height index (n-sn) there was significant relationship between experiment and control group which illustrated that nose height in patients with thalassemia is shorter while in other nose indices such as nose width (al-al) there was no significant difference. Although nasolabial angle showed considerable difference (3 degree) in thalassemia people, it was not significant statistically.
5. From facial indices, mandible height (sto-gn) showed maximum difference in compare to thalassemia groups which shows higher heath of mandible was significant statistically among thalassemia patients (thalassemia group: 48/7±3/25 and control group 5/125±41/74) and facial morphologic height indices (n-gn) and physiognomic height of upper part of face were reduced which indicates reduction of middle part of face. In addition, mandible depth (t-gn) and maxilla depth (t-sn) was not significant statistically $p<0.1$. Mandible width in-

Table 3. head parameters

Parameter name	thalassemia / landmark	Normal	suffered	P-value
Head height	(v-n)	105.3±5.7	96.99±7.28	P<0.001
Head round	(g-op)	562.5±3.45	560.23±2.73	P<0.01
Head length	Cirum (g-op)	190.55±5.85	185.46±4.86	P<0.001
Head width	(eu-eu)	155.25±4.65	154.41±2.92	P<0.4

Table 4. ear parameters

Parameter name	thalassemia / landmark	Normal	suffered	P-value
Earlobe width	(pra-pa)	35.7±2.6	35.15±2.70	P<0.4
Earlobe length	(sa-sba)	61.4±4.15	59.95±3.136	P<0.01

Table 5. eye parameters

Parameter name	thalassemia / landmark	Normal	Suffered	P-value
both inner corners of the eye	(en-en)	32.5±2.8	31.98±2.375	P<0.4
both outer corners of the eye	(ex-ex)	93.5±2.75	97.29±2.65	P<0.001
Eye gap length	(en-ex)	31.39±1.8	34.72±2.04	P<0.001

Table 6. nose parameters

Parameter name	thalassemia / landmark	Normal	suffered	P-value
nose width	(al-al)	34.75±2.35	33.85±2.35	P<0.1
Nose tip photogene	(sn-prn)	19.6±2.4	11.80±1.58	P<0.01
Nose height	(n-sn)	57.6±3.55	50.154±3.09	P<0.001
Nose bridge height	(bridge inc)	32.45±5.85	32.15±5.9	P<0.8
Nasolabial angle	Nasolabial angle	95.8±9.7	98.55±9.11	P<0.1

Table 7. facial parameters

Parameter name	thalassemia / landmark	Normal	suffered	P-value
Mandible width	(go-go)	104.45±5.8	107.40±2.86	P<0.1
Face morphologic height	(n-gn)	123.75±4.7	118.37±6.51	P<0.001
Physiognomic height of upper part	(n-sto)	77.65±4	74.106±3.08	P<0.001
Maxilla depth	(t-sn)	127.9±5.2	125.39±6.78	P<0.1
Mandible depth	(t-gn)	144.4±5.95	142.21±5.69	P<0.1
Mandible height	(sto-gn)	41.74±5.125	48.7±3.25	P<0.001
Face width	(zy-zy)	136.35±4.85	148.11±6.43	P<0.001

Table 8. on third of facial area

Parameter name	thalassemia / landmark	Normal	suffered	P-value
Physiognomic height of face	(tr-gn)	183.5±9.7	180.08±10.2	P<0.1
Forehead height	(tr-g)	56.55±6.35	57.32±4.45	P<0.5
Space between glabella and subnasal	(g.sn)	66±4.35	57.31±6.14	P<0.001
Lower height of face	(sn-gn)	66.83±5.7	68.55±4.2	P<0.1
Upper face profile inclination	(g-sn) inclination	2.55±3.4	8.67±6.037	P<0.001
lower face profile	(sn-pg) inclination	10.3±7.25	10.65±7.46	P<0.8

Table 9. upper lip parameter

Parameter name	thalassemia / landmark	Normal	Suffered	P-value
Mouth width	(ch-ch)	48.75±3.3	48.86±2.55	P<0.8
Upper lip height	(sn-sto)	21.1±2.4	23.40±5.24	P<0.02
height of cutaneous upper lip	(sn-ls)	14.1±2.55	21.73±3.75	P<0.001
Height of upper vermilion	(ls-sto)	7.3±1.6	6.8±1.42	P<0.2
Upper lip inclination	(sn-ls) inclination	7.25±8.95	12.03±9.302	P<0.05

dicator (go-go) was increased, it was not significant statistically (thalassemia group: 107/40±2/86 and control group 5/8±104/45). However facial width indicator was increased in these patients and showed significant difference in compare to control group.

6. Out of 1 third of facial indices, space between glabella and subnasal showed maximum difference in compare to thalassemia groups (thalassemia group: 57/31±6/14 and control group 66±4/35) which is indicator of decrease in one third of facial part in patients. Forehead height and lower height of face is indicator of decrease and physiognomic height of face was indicator of increase in patients with thalassemia but that was not significant statistically upper profile inclination indicated excrescence of subnasal in thalassemia patents and it was significant statistically. Moreover, difference in lower profile inclination of face was not significant in thalassemia patients and control group statistically P<0/8.

Indicators of mouth width (ch-ch) and height of upper vermilion (ls-sto) did not show significant difference in compare to control group. However, height of cutaneous upper lip (sn-ls) and upper lip height (sn-sto) indicated increased difference which were significant statistically. Out of them height of cutaneous upper lip showed maximum difference (thalassemia group: 21/73±3/75 and control group 14/1±2/55).

7. Out of lower lip indices, lower lip inclination (sl-li inclination) showed maximum difference and it was increased in patients but it was significant statistically (thalassemia group: 52/34±9/63 and control group 48/85±8/85) indices of lower vermilion height (sto-li) and height of lower lip was decreased out of these two difference, vermilion was significant p<0.001 (thalassemia group: 8/23±2/8

Table 10. lower lip parameters

Parameter name	thalassemia / landmark	Normal	suffered	P-value
lower lip height	(sto-sl)	18.05±2.05	17.27±2.84	P<0.2
cutaneous lower lip height	(li-sl)	9.25±1.85	9.61±2.89	P<0.5
lower vermilion height	(sto-li)	9.95±1.65	8.23±2.08	P<0.001
lower lip inclination	(sl-li) inclination	48.85±8.85	52.34±9.63	P<0.1

Table 11. chin and neck parameters

Parameter name	thalassemia / landmark	normal	Suffered	P-value
Upper height of chin	(sl-pg)	13.5±3.25	16.17±4.580	P<0.01
lower height of chin	(pg-gn)	19.3±2.9	20.55±2.751	P<0.001
height of chin	(sl-gn)	26.01±2.9	29.55±2.121	P<0.02
Neck length	(gn-c)	42.15±6.4	38.56±6.367	P<0.02
Mentocervical angle	Mento cervical	82.65±13.75	84.01±11.12	P<0.5

and control group 9/95±1/65). And height of cutaneous lower lip (li-sl) indicated significant difference between patients and control group.

8. Out of chin and neck indicators, chin indicators were all significant and they were increased from which upper height of chin (sl-pg) showed maximum difference (thalassemia group: 52/34±9/63 and control group 48/85±8/85) and also throat length (gn-c) was reduced in this indicators and that was significant and mentocervical angle was also ncreased however its statistical difference with control group was not significant p<0.5.

One main issue in treatment of jaw and facial abnormalities is accessing to anthropometric indicators of patients in a race group. This study aimed at analysis of the relationship between anthropometric sizes of head and facial soft tissue with major thalassemia and comparing them with Iranian adult's population. Previous studies mostly were performed on photography and cefalometry which were two dimensional documents of a three dimensional bodies and performed on hard tissue. In addition, there are many studies performed on identifying thalassemia effects on anthropometric sizes of head and facial soft tissue. Results of the study showed that anthropometric sizes of head and facial soft tissue in patient with control group (healthy people) were different. It is clear that obtained results can provide more accurate anticipation in changes of head tissue and so head and facial soft tissue and cause more precise orthodonsy of these people. At the following, we provide parameters of this abnormalities and obtained results.

37 linear indicators and 7 angle index were measured based on Method F directly on head and facial area. This study indicated that comparing both groups (control and thalassemia) proved that in one third of facial indicator significant difference happens. One of these changes is related to one third of lower part of face which includes increased mandible and chin height that is derived from vertical growth of mandible. In addition, in indicators related to one third of facial change we can have mentioned to space between glabella to subnosal and physiognomic height of upper part of face was decreased which generally shows size decrease of this part. Maxilla had anterior rotation which is anti-clock wise. Sule bassimitici researches of Dr Alavai et al and Dr Amini was compatible with our research results. In addition, in this study changes rate in indicators of ear were so inconsiderable which shows that effect of this disease is not considerable on cartilaginous tissue.

One other difference in head and facial area is related to nasal area that in these patients photogene of nose tip is was decreased. Nose height was reduced a lot too. As it is expected saddle form nose or in other words this rat looking face was justified with decrease of face middle third part and maxilla protogene. Results of this research were compatible with Mina and Pakshir.

In compare to control group, some angle change of face such as profile inclination of upper face and inclination of upper lip had been increased significantly (p<0/001). One of the most popular changes that happens in head and facial area of patients is pre maxilla excess which can be explained in relation with hyperplasia of bone marrow. According to this excess which is along with mandible vertical growth, rat looking face or leptospirosis with decrease on can be interpreted. Dr Amini researches also approved those results.

Many studies approve these findings; Including Seyedi and Nabavi Zadehin 2001 who studied jaw and oral and facial signs. These studies showed that 60/5% of lower jaw protrusion and 5% had saddle form nose. Mina and Pakshir also indicated that in patients with thalassemia some considerable changes such as skeleton relation class 2 and vertical growth of jaw happened.

In chin and neck zone, neck length throat (gn-c) happened significantly. In case of (g-op) index, head size has similar change which is approved by Mina and Pakshir studies. In addition, mandible and Zygomatic width are increased significantly which is approved by Alavi studies.

CONCLUSION

In patients with thalassemia, hyperplasia of bone marrow happens due to anemia so flat bones of face and head such as maxilla, Frontal and ramus mandible have lateral growth and change their involved soft tissue .

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