Cephalometric analysis of craniofacial deformity of β-thalassemic major by using computed tomography

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ABSTRACT

Background: Thalassemia is a hereditary anemia resulting from defects in hemoglobin production. β- thalassemia caused by decrease in the production of β- globin chains affect multiple organs and is associated with cranio-oro-facial deformity which include prominent cheek bones and protrusive premaxilla with depression of the nasal bridge often referred to as "rodent or chip-munk face" with small mandible and Cl.II skeletal relationship. This study aimed to investigate cephalometric craniofacial parameters (skeletal) of β- thalassemic major patients by using computed tomography and to compare findings with a group of healthy patients in the same age group.

Subject, Materials and Method: The study included (40) patients with β- thalassemic major (20 female and 20 male) with age 8-15years compared with (40) healthy controls (20 female and 20 male) with the same age, who admitted to spiral computed tomography scan unit in X-ray institute in AL-KARKH general hospital to have computed tomography scan for the brain, paranasal and for orthodontic purpose from October 2011 to June 2012. Cephalometric analysis of the selected four skeletal linear measurements and four skeletal angular measurements, by using direct analysis with software programs in a computer which is part of the computed tomography machine.

Results: There was no statistically significant difference between thalassemic males and females in all selected skeletal linear and angular measurements, Thalassemic patients have a highly significant large ANB angle and cl II skeletal relationship, significant larger gonial angle, Mandibular base length (Me-Go) is significantly shorter, Retrognathic mandible (SNB) is significantly decreased, highly significant shorter in total anterior facial height (N-Me) and total posterior facial height (S-Go), and also the Ramus height is highly significant decreased.

Conclusion: In thalassemic patients, the skeletal morphology is recognizable and mandible is retrognathism and they have skeletal cl. II pattern and Computed tomography is useful tool for assessment of the cranio facial measurement.

Key words: Thalassemia, computed tomography, Skeletal, Cephalometrics. (J Bagh Coll Dentistry 2013; 25(4):39-43).

INTRODUCTION

Thalassemia is blood dyscrasia characterized by a peculiar alteration in skull and long bone structures, which produce a "rodent facial appearance".[1]It is considered as heterogeneous inherited disorders that arise from mutation in the globin genes that reduce or totally abolish synthesis of one or more of the globin chains of hemoglobin[2] and produces a wide variety of signs and symptoms and complications in those who inherit this disease.[3]The modern concept of thalassemia is based upon studies carried out by many scientists; they classify it clinically into major, minor and intermedia depending on the presence or severity of the symptoms.[4]While the genetic classification is based on the globin chain involved into α and β-thalassemia.[5]The thalassemic patients characterized by having a prominent frontal bossing and bulging of the forehead and prominent frontal and parietal bones.[6]

In the skull of older child; radiographically shows reformation of outer table in the frontal bone and resolving process in the parietal bone.

Thalassemic patient have high malar eminence and high bulging cheek bones, which give rodent features and the prominent malar eminence producing obvious malocclusion in these patients.[6]

Bi-maxillary expansion is one of the classical clinical changes and the progressive maxillary enlargement, maxillary dysplasia, sever protrusion of the middle third of the face and maxillary tuberosity become widened; all these changes producing the typical facial appearance with sever degree of malocclusion.[7]

Many authors [8] found posterior rotation of the mandible and he found posterior (clock wise) mandibular rotation in the thalassemic children, the mandible have thin cortex and trabecular appear coarse in pattern with enlarged marrow space which is depressed as "chicken–wire".[9]

Dentition shows protrusion, flaring and spacing maxillary anterior teeth, open bite and other type of malocclusion [9, 10] as in figure (1).

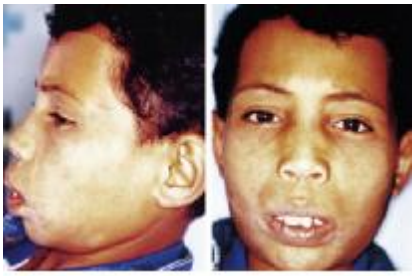

Figure 1. Photographic picture shows the facial appearance and Malocclusion in beta thalassemic patient

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SUBJECTS, MATERIALS AND METHODS

A prospective study consists of (80) patients ranged from (8-15) years, divided into two groups, thalassemic patients and health group, each group composed of (40) subjects (20 males & 20 females) who admitted to spiral computed tomography scan unit in X-ray institute at AL-KARKH general hospital in Baghdad to have computed tomographic scan for the brain, paranasal and for different orthodontic purposes (both for thalassemic patients and control groups) from October 2011 to June 2012.

Patient selected with no history of orthodontic treatment and maxillofacial surgery and facial trauma, no facial asymmetry and Class-I molar relationship.

The control group has the same criteria but in addition to: healthy and had no disease of genetic origin, no gross skeletal defects,[11] Bilateral Cl. I molar relationship based on angle classification normal over get and over bite(2mm), Very mild spacing (accepted) or crowding (0-1) mm.[12]

The examination was performed on multi–slice spiral tomography scanner (The Philips Brilliance CT-64 Thickness of slice 0.5m).

Method

Cephalometric analysis:

1-3D measurements were measured by distance and angular tools of commercial software. Each anatomic measurement was identified as a 3D point with the software.

The software enables simultaneously recognizing the same spatial point in sagittal, coronal and axial planes, which are represented in three separate windows. A fourth window allows the recognition of the anatomic point on a volume rendered (VR) window, which is a 3D image of the skull (in sagittal view).[13]

Cephalometric points or anatomic landmarks were identified on its position by using definition of each point or landmark and draw line and angle between them (as in figure 2), using a sagittal view of the volume rendered window and measure the linear and angular measurements on the software of the CT scan.

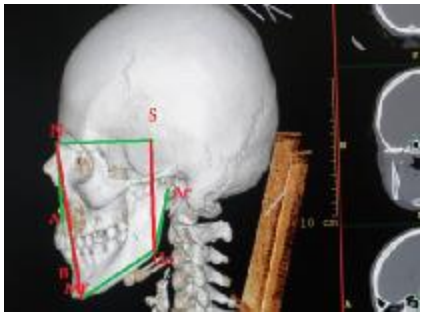

Figure 2. Volume rendered (VR) window with the linear and angular measurements on the software of the CT scan.

Cephalometric Planes:

Mandibular plane (GO. Me), Anterior facial height (N,Me), Posterior facial height (S.Go), Ramus height (Ar-Go).

Angular Measurements:

Ar. Go. Me (cephalometric gonial angle), SNA (Skeletal), SNB angle (skeletal), ANB angle (skeletal).

RESULTS

- **Case (Thalassemia) - control difference in mean of skeletal linear measurements:** The mean linear distance (Go-Me) was significantly lower in thalassemic patients (67.6±7.7) mm compared to control (75.3±7.7) mm. The mean linear distance (Ar-Go) was significantly lower in thalassemia patient (41.6±3.3) mm compared to control (44.9±3.3) mm. The mean linear distance (S-Go) was significantly lower in thalassemic patient (69.1±6.3)mm compared to control (75.4±6.3)mm, and the mean linear distance (N-Me)was significantly decreased in thalassemic patient (116.5±6.7)mm compared to control (123.2±6.7)mm. Table (1)& Figure(3).
- **Case (Thalassemia)- control difference in mean of skeletal angular measurements:** The mean of angular distance (Ar-Go-Me) was significantly higher in thalassemia (133±4) mm compared to controls (129±4) mm, the mean of the angle (SNA) no significant affected because the mean value in thalassemia (80.5±0.4) mm and in control (80.1±0.4) mm, the mean of the angle (SNB) was significantly lower in thalassemia (73.1±3) mm compared to controls (76.1±3) mm, the mean of the angle (ANB) was significantly increase in thalassemia (7.6±3.2) mm compared to controls (2.4±3.2) mm. Table(2) & Figure (4)

Table 1: The case-control differences in mean of selected linear measurements.

Measurements	Controls	Cases Thalassemia	P (t-test)	Difference in mean
Angle (Ar_Go_Me)				
Range	128-136	128-138.1		
Mean	129	133	<0.001	4
SD	1.4	2.7		
SE	0.22	0.43		
N	40	40		
Angle SNA				
Range	72-87	76-84		
Mean	80.1	80.5	0.4[NS]	0.4
SD	2.3	1.9		
SE	0.37	0.3		
N	40	40		
Angle SNB				
Range	70.8-78	70-76		
Mean	76.1	73.1	<0.001	-3
SD	2.1	1.5		
SE	0.34	0.24		
N	40	40		
Angle ANB				
Range	1-10.4	3-12.8		
Mean	4.4	7.6	<0.001	3.2
SD	2.7	2.2		
SE	0.42	0.35		
N	40	40		

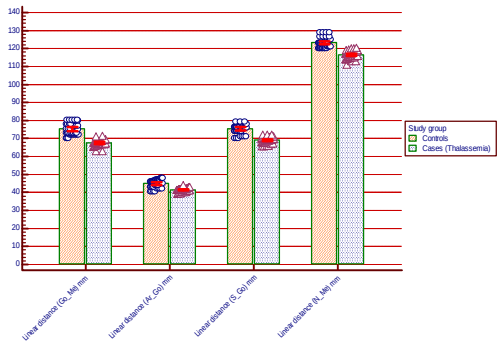

Figure 3. Dot diagram with error bars showing thecase-control difference in mean (with its 95% confidence interval) of selected linear measurements.

Table 2: The case-control difference in mean of selected angular measurements.

Linear distance (Go_Me) mm	Partial regression coefficient	P
(Constant)	68.6	<0.001
Age in years	0.60	<0.001
Male gender compared to female	-1.05	0.06[NS]
Having thalassemia compared to controls	-7.50	<0.001
Linear distance (Ar_Go) mm	Partial regression coefficient	P
(Constant)	40.7	<0.001
Age in years	0.34	<0.001
Male gender compared to female	0.22	0.56[NS]
Having thalassemia compared to controls	-3.20	<0.001
Linear distance (S_Go) mm	Partial regression coefficient	P
(Constant)	72.6	<0.001
Age in years	0.21	0.06[NS]
Male gender compared to female	0.63	0.21[NS]
Having thalassemia compared to controls	-6.29	<0.001
Linear distance (N_Me) mm	Partial regression coefficient	P
(Constant)	118.9	<0.001
Age in years	0.31	0.013
Male gender compared to female	1.33	0.016
Having thalassemia compared to controls	-6.55	<0.001

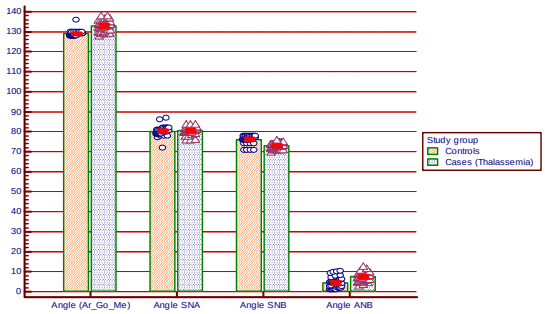

Figure 4. Dot diagram with error bars showing the case-control difference in mean of selected angular measurements.

Table 4: Effect of age and gender on skeletal angular measurements by using multiple linear regressions.

Angle (Ar_Go_Me)	Partial regression coefficient	P
(Constant)	123.6	<0.001
Age in years	0.47	<0.001
Male gender compared to female	-0.43	0.33[NS]
Having thalassemia compared to controls	4.19	<0.001
Angle SNA	Partial regression coefficient	P
(Constant)	76.3	<0.001
Age in years	0.34	0.001
Male gender compared to female	-0.43	0.35[NS]
Having thalassemia compared to controls	0.52	0.24[NS]
Angle SNB	Partial regression coefficient	P
(Constant)	71.8	<0.001
Age in years	0.37	<0.001
Male gender compared to female	-0.24	0.53[NS]
Having thalassemia compared to controls	-2.87	<0.001
Angle ANB	Partial regression coefficient	P
(Constant)	4.5	0.005
Age in years	0.01	0.95[NS]
Male gender compared to female	-0.39	0.49[NS]
Having thalassemia compared to controls	3.15	<0.001

DISCUSSION

The result show that all selected skeletal linear and angular measurements are not significantly different of level (p>0.05) for both thalassemic males and females. These findings come in agreement with the findings of Bassimitci, Abu–AL-Haija [8,14] and Moutaz [10]who found that the same results that thalassemic males and females posses no significant differences.

It is indicate statistically no significant difference in SNA angle between thalassemic (80.5±0.4mm) and controls (80.1±0.4mm). The finding comes in agreement with the findings of Bassimitci, Abu–AL Haija[8,14] and Moutaz [10]who found that there were insignificant increase in tendency to sagittal maxillary over growth was observed and the anterio-posterior position of the maxilla in relative to cranial base are not different indicating that other variables are responsible for the occurrence of discrepancy, there is statistically significant reduction in SNB angle in thalassemic patient (73.1±3mm) compared to controls (76.1±3mm). This finding coincides with agreement with Moutaz [10] and disagreement with Bassimitci [8] and Abu–AL Haija [14]according to these findings the thalassemic patients exhibited significantly retrognathia in the mandible, ANB Angle highly significant increased in thalassemic patients (7.6±3.2mm) compared to controls (2.4±3.2mm). (Normal limit 2-4mm).This finding coincides with the findings of Bassimitci [8] and Abu–AL Haija[14] and Moutaz[10]. According to this finding the thalassemic patients exhibited a large inter-maxillary discrepancy since that the ANB angle produced CL11 skeletal pattern, there is a significant increase in gonial angle which is in thalassemic patients (133±4mm) compared to controls (129±3mm). This finding comes in agreements with the findings of Bassimitci[8] and Moutaz[10], This increase in the measurement of gonial angle indicates more of a tendency to posterior rotation of the mandible with condylar growth directed posteriorly. The mandibular body length (Me-Go) is significantly decreased in thalassemic patients (67.6±7.7mm) compared to controls (75.3±7.7mm). These findings come with agreement with Bassimitci [8] and Abu–AL Haija[14]and Moutaz [10].

The results indicate that the mandible is unusually short, there is a highly significant difference with shorter total anterior facial height for thalassemic patients (116.5±6.7mm) compared to controls (123.2±6.7mm).

This finding comes with agreement of Moutaz [10] and disagreement with Bassimitci [8] and Abu–AL Haija [14] who found that thalassemic patients have a shorter total anterior facial height with no significant difference, the total posterior facial height significant decreased in thalassemic patients (67.6±7.7mm) compared to controls (75.3±7.7mm). This finding coincides with Bassimitci, Abu–AL Haija [8,14], and Moutaz [10]; found that thalassemic patients possess shorter posterior facial height but is not significantly different. The posterior facial height is largely determined by growth at the condyle, which is deficient probably due to anemia.

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