



Dental Arch Dimensions in Subjects with Beta-thalassemia Major

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ABSTRACT

Aim: Thalassemia is a group of inherited hemoglobinopathies with thalassemia major representing the severe form of the disease characterized by craniofacial deformities. The aim of this study is to provide a detailed description of dental arch dimensions in subjects with thalassemia major.

Subjects and methods: The sample consisted of 43 thalassemic subjects, 24 males and 19 females, aged 7.3 to 15.4 years (mean \pm SD = 10.6 \pm 3.5 years) and control group matched by age and sex. Dental casts of the participants were measured for arch lengths, arch widths, using a digital sliding caliper. Student t-test was used for comparison of mean values between males and females as well as between thalassemic and control groups.

Results: The results show that all means of maxillary and mandibular arch dimensions in thalassemic males and females were smaller than their controls, with 14 of the 16 comparisons being statistically significant (ranged from $p < 0.05$ to $p < 0.001$). The segmental arch lengths in the maxilla and mandible of thalassemic group were reduced by an average of 2.59 and 2.55 mm respectively, compared with the control group. The mean maxillary and mandibular arch depths (lengths) in the thalassemic group were shorter by 3.21 and 2.63 mm respectively, relative to the controls ($p < 0.001$). All arch widths thalassemic patients were significantly reduced by an average ranged from 1.33 to 1.90 mm in the maxilla and 1.37 to 1.77 mm in the mandible.

Conclusion: The present study showed that the maxillary and mandibular dental arches dimensions are significantly reduced in patients with thalassemia major compared with healthy control subjects.

Clinical significance: Changes in the size of dental arches and tooth dimensions in thalassemic patients have an impact on the occlusal relationships. These changes should be taken into account when planning orthodontic treatment and orthognathic surgery.

Keywords: Dental arch, Dimensions, Thalassemia major.

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INTRODUCTION

Thalassemia refers to a group of hemolytic anemia involving inherited defects in the synthesis of either alpha or beta-polypeptide chains of hemoglobin, leading to decreased hemoglobin production and hypochromic microcytic anemia. Because of this defect, the condition referred to as alpha or beta-thalassemia, with several subtypes manifested in diverse clinical pictures. Based on genetic heterogeneity and clinical and hematological variability, thalassemia is classified as homozygous, heterozygous, or compound heterozygous. The homozygous type of beta-thalassemia (also known as thalassemia major, Cooley's anemia or Mediterranean anemia) exhibits the most severe clinical symptoms, often described as transfusion-dependent disorder.

Thalassemia is considered among the most common genetic disorder worldwide, presenting major public health and social problems in the high incidence areas. About 3% of the world's population carry beta-thalassemia gene. The disorder is most common among individuals of Mediterranean particularly of Southern Italy, Greece and Cyprus with prevalence 10 to 15%. The condition is also described in Arab countries, Turkey, Iran, Southeast Asia and Africa, with frequency ranged between 1.5 to 5%.^{1,2} In North America, thalassemia is noted primarily in persons of Italian and Greek decent and in blacks.

Beta-thalassemia major is life-threatening condition commonly manifested during early infancy. Thereafter, affected children become progressively pallor, severely anemic and fail to thrive. They suffer from feeding problems, recurrent fever, bleeding tendencies especially toward epistaxis, susceptibility to infection, pathological fractures of long bones and vertebrae, osteoporosis, endocrine abnormalities, splenomegaly, lack of sexual maturation and retardation of growth.^{1,2} Patients with beta-thalassemia usually require blood transfusion to keep their hemoglobin level near normal in order to abate the symptoms of hypoxia.

Some of the most striking changes in thalassemia major are seen in orofacial expressions and craniofacial deformities. Skeletal abnormalities result primarily from hypertrophy and expansion of the erythroid marrow consequent to ineffective erythropoiesis. In severe cases the facial appearance includes prominent frontal and cheek bossing, depression of the bridge of the nose, protrusive premaxilla (chipmunk facies, Fig. 1), flaring of the maxillary anterior teeth, lip retraction and varying degrees of malocclusion.³⁻⁵ Thalassemia subjects are also at high-risk of dental caries and periodontal disease.⁶⁻⁸



Fig. 1: A 14-year-old girl showing typical facial features of thalassemia major. Note: the prominent frontal and cheek bosses, saddle nose and protrusive premaxilla (chipmunk facies)

Little data on the dental arch morphology are available on this widely distributed disorder. The aim of the present study was to determine dental arch dimensions and their clinical applications in Jordanian subjects with thalassemia major and to compare the result with unaffected (thalassemia-free) control group.

MATERIALS AND METHODS

The sample comprised of 43 patients with thalassemia major, 24 males and 19 females aged 7.3 to 15.4 years, with the mean age (\pm standard deviation) of 10.6 ± 3.5 years. Unaffected control group matched by age and sex to the study group were tested. Family history revealed that 41% of the patients were the product of first-cousin marriage, 32% of second-degree cousins, and 27% of distally related or not related. Physical growth assessment carried out on 54 thalassemic patients aged between 5.5 and 18.3 years showed that 42.1% of males and 35.7% of females were below the third percentile in height. The weight of 57.8% of

the males and 41.7% of females were below the third percentile, on the growth chart of Jordanian school children.

Alginate impressions were taken in suitable perforated trays for the maxillary and mandibular dental arches of every subject, and cast immediately in dental stone. Measurements of arch dimensions were made on the casts using fine tips electronic digital sliding caliper reading to 0.1 mm. Arch perimeters were measured using a brass wire contoured along the line of occlusion and then straightened out. The following measurements (in mm) were conducted:

- *Arch depth:* Distance from the line connecting the distal surface of the first molars to the midpoint between the central incisors (Fig. 2).
- *Anterior arch length:* Distance between the mesial contact point of the central incisor and the distal contact point of the canine (Fig. 2).
- *Posterior arch length:* Distance between the mesial contact point of the canine and the distal contact of the second premolar (Fig. 2).
- *Interincisor width:* Distance between the distal contact point of the lateral incisor on one side to the distal contact point of the contralateral tooth (Fig. 3).
- *Inter canine width:* Distance between the cusp tip of the canine on one side to the cusp tip of the contralateral canine (Fig. 3).
- *Interpremolar width:* Distance between the buccal cusp tip of the second premolar on one side to the buccal cusp tip of the contralateral second premolar (Fig. 3).
- *Intermolar width:* Distance between the mesiobuccal cusp tip of the first molar on one side to the mesiobuccal cusp tip of the contralateral first molar (Fig. 3).
- Arch perimeter was measured as a line from the distal aspect of the first molar passing around the arch on the contact points and incisal edges to the distal aspect of the contralateral first molar (Fig. 2).

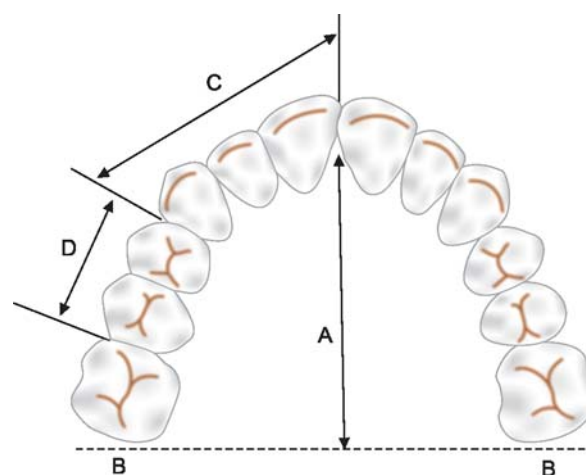


Fig. 2: Measurements recorded: A – arch depth; B – arch perimeter; C – anterior arch length; D – posterior arch length

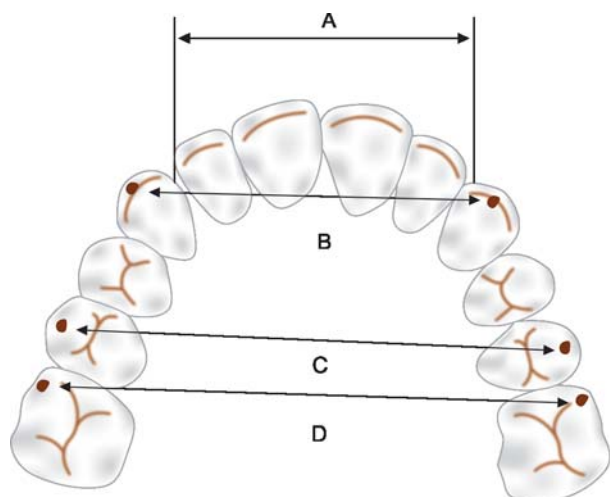


Fig. 3: Arch width measurements: A – Interincisor width; B – Inter canine width; C – interpremolar width; D – intermolar width

Descriptive statistics including the mean of arch length, arch width, and arch perimeter, standard deviation (SD), standard error of the mean (SEM), and coefficient of variation ($CV=100\text{ SD}/\text{mean}$) were computed. The t-test for independent samples was performed for comparison of mean values between males and females as well as between thalassemic group and control group.

RESULTS

Dental arch dimensions for thalassemic and control groups are given in the Table 1. The segmental arch lengths (the sum of the anterior and posterior arch lengths) in the maxilla and mandible of the thalassemic group were reduced by an

average of 2.59 and 2.55 mm respectively, compared with the control group. No significant differences were found in the arch lengths between the right and left side of the dental arches. The arch depths of the maxilla and mandible in thalassemic group were shorter than the controls by 3.21 and 2.63 mm, respectively. These differences were statistically significant ($p < 0.001$). The anterior arch lengths in the thalassemic group were 1.76 mm shorter in the maxilla and 1.63 mm shorter in the mandible compared with the control group. The differences between the two groups were statistically significant ($p < 0.05$ and $p < 0.001$ respectively). There was a tendency toward shorter posterior arch length in the maxillary (0.83 mm) and the mandible (0.92 mm), but it did not reach the significant level of $p < 0.05$.

The differences in dental arch widths between thalassemic group and control group ranged from 1.33 to 2.19 mm in the maxilla and 1.15 to 1.77 mm in the mandible. The differences between the two groups were statistically significant ranged from $p < 0.05$ to $p < 0.001$ (t-value ranged from 2.14 to 4.79, degree of freedom = 69 to 78). The interincisal widths of both arches ranked the highest order of reduction compared with controls ($p < 0.001$), measured 1.82 mm in the maxilla and 1.70 mm in the mandible. The mean maxillary and mandibular arch perimeters were reduced by 3.91 and 3.44 mm respectively, in the thalassemic group relative to the controls ($p < 0.001$). The intermolar widths in thalassemic group were reduced by 2.19 and 1.77 mm in the maxillary and mandibular arches respectively. These differences were statistically significant ($p < 0.01$).

Table 1: Means and standard deviations (in mm) of dental arch dimensions in thalassemic and control groups (sexes pooled)

Parameter	Thalassemia		Control		Diff [†]
	Mean	SD	Mean	SD	
Maxilla					
Arch depth	38.66	2.53	41.87	2.74	3.21***
Anterior arch length	22.03	1.94	23.79	2.06	1.76*
Posterior arch length	14.10	1.76	14.93	1.87	0.83
Interincisal width	27.04	2.08	28.86	1.95	1.82***
Inter canine width	33.09	2.44	34.42	2.11	1.33*
Interpremolar width	45.19	2.56	46.53	2.63	1.34*
Intermolar width	49.41	2.73	51.60	2.59	2.19**
Arch perimeter	95.83	3.96	99.74	3.81	3.91***
Mandible					
Arch depth	33.59	2.54	36.22	2.76	2.63***
Anterior arch length	16.74	1.63	18.37	1.54	1.63***
Posterior arch length	13.61	1.81	14.53	1.82	0.92
Interincisal width	20.92	1.73	22.62	2.04	1.70***
Inter canine width	26.43	2.05	27.58	1.97	1.15*
Interpremolar width	39.66	2.59	41.34	2.68	1.68**
Intermolar width	44.95	2.76	46.72	2.93	1.77**
Arch perimeter	86.87	3.61	90.31	3.75	3.44***

Diff[†] = differences between means

* $p < 0.05$, ** $p < 0.01$, *** $p < 0.001$

DISCUSSION

Tooth crown size and dental arch dimensions are required in planning orthodontic treatment. Discrepancy between tooth size and arch length result in dental crowding;^{9,10} the most common type of malocclusion.¹¹ A relationship has also been demonstrated between tooth size and arch dimension in third molar eruption and impaction.^{12,13}

Dental arch size and shape are influenced by a variety of factors including genetic, environmental, pathological conditions, eruption; position, number of teeth, and ethnic diversity.^{11,14-17} The present study showed that all arch dimensions, were reduced in the thalassemic group compared to the controls. In 14 of the 16 comparisons, the differences between the two groups were statistically significant. In thalassemic subjects, the arch length was 3.21 mm shorter in the maxilla and 2.63 mm shorter in the mandible compared to the controls. The mandibular arch depth in thalassemic subjects showed less difference in dimension than those in the maxilla when compared to the controls (Table 1). This could be related to the dense cortical plates of the mandible with apparently prevention of the expansion.

The present study showed that the mean maxillary and mandibular depths in thalassemic patients were significantly shorter than in the control group. A review of the literature reveals some inconsistency regarding the interrelation between the maxilla and the mandible in thalassemia. A previous study showed that about one-fourth of 54 thalassemic patients exhibited protrusion of the maxillary incisors and increased overjet and 17% displayed gross craniofacial deformity⁸ (Figs 4 and 5). The prominence of the premaxilla in severe cases has led some authors to conclude that the maxillary incisors were proclined.^{18,19} However, cephalometric analysis of thalassemic patients demonstrated that the maxilla appeared prominent due to a reduced cranial base length associated with smaller and retruded mandible resulted in class II malocclusion^{5,20} (Fig. 5). Evidence indicated that the manifestations of thalassemia major depend on the severity of the anemia, the patient's age, the duration of the clinical symptoms, the timing and frequency of blood transfusion, age of initiation of iron chelation therapy, bone marrow transplantation and splenectomy.

The present findings showed that thalassemic patients had growth retardation with 42.1% of the males and 35.7% of females who were below the third percentile in height. The growth retardation was more apparent in the late childhood and increased with age. In this context, Lapatsanis et al²¹ showed that in five to seven years old thalassemic children half of them had bone retardation (>6 months),

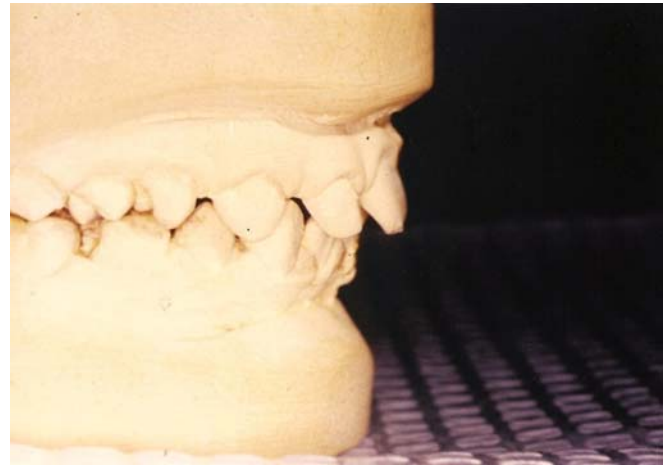


Fig. 4: Dental casts of thalassemic patient showing flaring and spacing of the maxillary anterior teeth and malocclusion



Fig. 5: Lateral cephalometric radiograph of a 13-year-old male with thalassemia major. The prominent premaxilla caused drifting of the incisors and excessive overjet. *Note:* Class II malocclusion and vertical growth direction

whereas after this age retardation was found in almost two-third of the subjects. Parallel with these findings, craniofacial deformities in thalassemic patients become more apparent in the older age group.^{4,5,8} The shorter arch depth and length and the narrower arch width in this study could be a reflection of general growth retardation and skeletal changes in thalassemic patients. In addition, the reduced tooth size in this disorder²² may render the dentoalveolar bone housing the teeth to be more deficient. Evidence supporting the relation between reduced tooth size and smaller dental arches has come from studies in patients with Down's syndrome,¹⁶ oligodontia,^{17,23} and cleft lip and palate.²⁴

CONCLUSION

Changes in the size of dental arches in thalassemic patients have an impact on the occlusal relationships. The present

study indicates that the maxillary and mandibular dental arches dimensions are significantly reduced in patients with thalassemia major compared with healthy control subjects. These findings should be taken into account when planning orthodontic treatment and surgical correction of maxillary deformities.

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