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Malocclusion among Sudanese Children with Beta-Thalassemia Major

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Abstract

Background: Thalassemias are inherited blood disorder, widespread amongst Italian, Middle Eastern, Greek, South Asian and African children. It is classified into Alpha and Beta Thalassemias with a number of subtypes. β-thalassemia major is the most sever one. The objective of this study was to determine the association between β-thalassemia major and malocclusion among a sample of Sudanese children compared to healthy children. *Method:* The present study was conducted for 54 children with β-thalassemia major aged (4-16) years old compared to 54 healthy control group matched with age and gender in Khartoum state, Sudan. Malocclusion was evaluated according to Angle's classification. Statistical analysis was done using the statically package for social science (SPSS) version 20. The 95% confidence intervals for the association between variables were estimated and compared using Chi-square test. p value ≤ 0.05 was considered statically significant. *Results:* β-thalassemia major exhibited various types of malocclusion; open bite (11.3%), increased overjet (37.7%), maxillary spacing (44.4%), maxillary crowding (16.7%), mandibular spacing (25.9%), and mandibular crowding (24.1%). Angle's class I was the most frequent occlusal relation (80.0%) in children with β-thalassemia major, (20.0%) had class II occlusal relation, and no one had class III. No statically significant difference between the two study groups in relation to all types of malocclusion except for overjet (p≤0.004). *Conclusion:* Various types of malocclusion were found in β-thalassemia major children in Sudan, increased overjet is the most frequent followed by anterior openbite. A significant correlation existed between β-thalassemia major and healthy control group. Perception of the situation of β-thalassemia children should contribute for the planning of dental services that can help in prevention of malocclusion and avoid future dental problems.

Keywords: β-Thalassemia Major; Malocclusion; Control Children.

Introduction

Haemoglobinopathies are inherited disorders of globin, the protein component of hemoglobin (Hb). Common Haemoglobinopathies are thalassemia and sickle cell disease, primarily characteristics of the tropic and sub tropic regions but are now globally wide spread due to migration [1,2].

Thalassemia is defined as an autosomal recessive inherited blood disorder, caused by defects in the synthesis of either Alpha or Beta globin chains, leading to hypochromic microcytic anaemia and decrease hemoglobin production [3,4].

Thalassemia is one of the most widespread genetic disorders and it was first discovered clinically by Dr. Thomas Cooley in 1925 [5,6].

Globally it has been estimated that 7% of the world population are carriers of hemoglobin disorders, and every year over 330,000 of born infants have disorders, 17% of them are thalassemia [2,7].

Worldwide thalassemia is more common among Mediterranean roots and origin population, predominantly in Italy, Greece, and Cyprus with a prevalence of 10% to 15%. Low prevalence 1.5–5% is reported in Arabian population; Turkey, South East Asia, Africa and Iran [8]. Moreover in India It is a major health concern, the prevalence of β-thalassemia is as high as 17% [9].

The first clinical manifestations of β-thalassemia major appear at 4-6 month of age as severe anaemia, feeding difficulties, growth retardation and failure to thrive [5].

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The clinical features of β-thalassemia major are characterized by bone marrow hyperplasia, skeletal deformities, hepatospleenomegaly and iron overload as a result of repeated blood transfusion [10,11]. Affected children are susceptible to foliate deficiency, cardiac failure and infection. It can affect the endocrine system in some cases [5,12-15].

The dentofacial abnormalities in β-thalassemia patients has various forms; protrusion of anterior teeth, spacing of teeth, increased over jet, depressed nasal bridge with enlargement of the maxilla and varying degrees of malocclusion which reflect characteristic facial appearance known as chipmunk appearance [5,16,17].

Diagnosis of thalassemia; by full blood count, hematological tests and molecular genetic analysis [1,18]

β-thalassemia major can be life threatening if left untreated before the age of three years (2,14). Nowadays prognosis has improved, and life expectancy is approaching normal as a result of medical advances in transfusion, iron chelation therapy and bone marrow transplantation [2,7,12].

In previous literature few studies are available concerning β-thalassemia and its association with dental caries and malocclusion [14]. To our knowledge no such study is available among Sudanese population. Therefore, the present study is designed to assess the association between β-thalassemia major, dental caries and malocclusion among a sample of Sudanese children and compare it with healthy children matching with number and age group.

Research hypothesis

Null hypothesis;

There is no association between β-thalassemia major, dental caries and malocclusion among Sudanese children.

Methodology

This study is an analytical cross-sectional carried out in 54 children with β-thalassemia major aged (4-16) years old compared to 54 healthy control group matched with age and gender.

The β-thalassemia:

Patients with β-thalassemia major attending Jaafar Ibn Oaf hospital, Al Buluk pediatric hospital, Mohamed Alamin Hamid pediatric hospital, Al Ban Jadeed hospital from March 2017 – July 2017 were examined after obtaining a written permission from the directors of the hospitals and after receiving an informed written consent from their parents.

All the β-thalassemia major patients who met the inclusion criteria were examined based on the availability of the patients at the four public hospitals, taking advantage of the scheduling of medical appointments and routine tests.

The healthy control group:

Multi stage sampling was applied; the first stage schools were selected from the same catchment area of the referral center, the second stage children in each school were selected following stratification by age and gender and the third stage children in each class were selected following systematic randomization technique.

Examination of children was carried out after a request letter explaining the purpose of the study was given to the responsible school authorities to carry out the research and after a written consent was received back from their parents.

Examination of the β-thalassemia patients and the healthy control group was done in an ordinary chair in a doctor's office and teacher's office respectively under artificial light. Before dental examination children were instructed to rinse there mouth for better visualization. Teeth were cleaned with cotton roll whenever necessary. The child was placed in an up-right position and the occlusion was assessed while the child was biting in maximal intercuspation. A note taker recorded the information as reported by the dentist during the examination.

An ethical approval was obtained from the research committee of the Faculty of Dentistry-University of Khartoum prior to the conduction of the study.

Malocclusion was recorded according to Bjork and Solow registration of malocclusion [19].

Statistical analysis:

Data was collected, cleaned and analyzed using the statically package for social science (SPSS) version 20. The 95% confidence interval for the association between dental caries was estimated and compared using Chi-square test. A level of significance of 5% will be adopted to reject the null hypothesis.

Results

A total of 108 children (70 boys, 38 girls) 4-16 years old were examined in this study.

The sample comprised of two equal groups; 54 patients with β-thalassemia major and 54 healthy group similar in age and sex distribution. The mean age for β-thalassemia major and for the healthy control group was 7.7±3.4 years. It was clear that the majority of the sample was boys, Table 1. According to the age the study sample was divided into three age groups, and the majority of them were in the age group 4-6 years old, Figure 1.

Age group 4-6 years old:

Table 2 showed the distribution of malocclusion among 4-6 years old children with β-thalassemia major and the healthy control children. All of the findings were insignificant.

Table 1: The distribution of the study sample according to gender (%)

Children	Boys	Girls	Total
β-thalassemia patients	35 (64.8)	19 (35.2)	54 (100)
Healthy Control children	35 (64.8)	19 (35.2)	54 (100)
Total	70 (64.8)	38 (35.2)	108 (100.0)

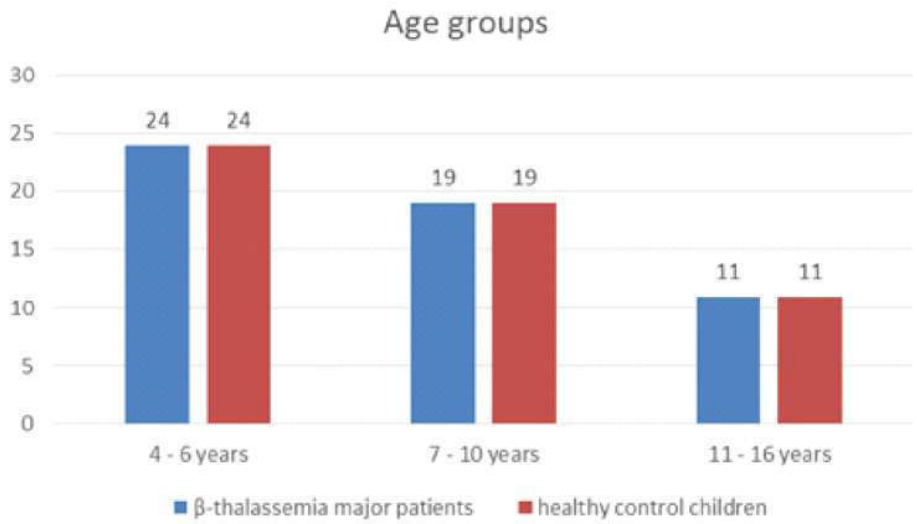

Fig. 1: Distribution of the study sample according to age group

Table 2: Distribution of malocclusion in the age group 4-6 years old children in the study sample (%).

Variables		β-thalassemia patients	Healthy Control children	P value
		N (%)	N (%)	
Terminal Plane	Mesial step	19 (79.2)	21 (87.5)	0.192
	Flush	2 (8.3)	3 (12.5)	
	Distal step	3 (12.5)	0 (0.0)	
Horizontal Overjet	Normal	16 (69.6)	21 (87.5)	0.313
	Increased	4 (17.4)	2 (8.3)	
	Decreased	3 (13)	1 (4.2)	
Vertical Overbite	Normal	21 (91.3)	22 (91.7)	0.513
	Increased	1 (4.3)	2 (8.3)	
	Anterior open bite	1 (4.3)	0 (0.0)	
Spacing in Maxilla	Normal	9 (37.5)	9 (37.5)	0.831
	Spaced	14 (58.3)	13 (54.2)	
	Crowded	1 (4.2)	2 (8.3)	
Spacing in Mandible	Normal	13 (54.2)	14 (58.3)	0.457
	Spaced	10 (41.7)	7 (29.2)	
	Crowded	1 (4.2)	3 (12.5)	

p value significant ≤0.05

Age group 7-10 years

Table 3 showed the distribution of malocclusion in the study sample at 7-10 years old. It was clear that the majority of the two groups had Angle's class II malocclusion and no one recorded with class III malocclusion.

Increased horizontal overjet occurred more among the β-thalassemia group, three cases had anterior open bite and the difference was not significant.

Crowding was recorded more in the mandible than the maxilla. A significant difference was recorded p=0.02 when the two study groups were compared.

Age group 11-16 years old

In table 4 the children with β-thalassemia major in the age group eleven to sixteen years old showed more increase in the prevalence of Angle's class II (36.4%), increased horizontal over jet (81.8%), anterior open bite (18.2%), crowded maxilla (27.3%) and crowded mandible (36.4%). A statistical significant correlation was observed in relation to horizontal overjet between the β-thalassemia major patients and the healthy control group p = 0.003.

Table 3: Distribution of malocclusion in the age group 7-10 years old children in the study sample (%)

Variables		β-thalassemia patients	Healthy Control children	P value
		N (%)	N (%)	
Angle's Classification	Class I	17 (89.5)	15 (78.9)	0.66
	Class II	2 (10.5)	4 (21.1)	
	Class III	0 (0)	0 (0)	
Horizontal Overjet	Normal	9 (47.4)	13 (68.4)	0.312
	Increased	7 (36.8)	3 (15.8)	
	Decreased	3 (15.8)	3 (15.8)	
Vertical Overbite	Normal	15 (78.9)	17 (89.5)	0.177
	Increased	1(5.3)	2 (10.5)	
	Anterior open bite	3 (15.8)	0 (0)	
Spacing of Maxilla	Normal	6 (31.6)	7 (36.8)	0.200
	Spaced	8 (42.1)	11 (57.9)	
	Crowded	5 (26.3)	1 (5.3)	
Spacing of Mandible	Normal	9 (47.4)	12 (63.2)	0.02**
	Spaced	2 (10.5)	6 (31.6)	
	Crowded	8 (42.1)	1 (5.3)	

p value significant ≤0.05

Table 4: Distribution of malocclusion in the age group 11-16 years old children in the study sample (%)

Variables		β-thalassemia patients	Healthy Control children	P value
		N (%)	N (%)	
Angel's Classification	Class I	7(63.6)	10(90.9)	0.311
	Class II	4(36.4)	1 (9.1)	
	Class III	0 (0)	0 (0)	
Horizontal Overjet	Normal	2(18.2)	9 (81.8)	0.003**
	Increased	9(81.8)	1 (9.1)	
	Decreased	0 (0)	1 (9.1)	
Vertical Overbite	Normal	9(81.8)	9 (81.8)	0.513
	Increased	0 (0)	1 (9.1)	
	Anterior open bite	2(18.2)	1 (9.1)	
Spacing in Maxilla	Normal	6(54.5)	7 (63.6)	0.528
	Spaced	2(18.2)	3 (27.3)	
	Crowded	3(27.3)	1 (9.1)	
Spacing in Mandible	Normal	5(45.5)	9 (81.8)	0.192
	Spaced	2(18.2)	1 (9.1)	
	Crowded	4(36.4)	1 (9.1)	

p value significant ≤0.05

Table 5: Distribution of malocclusion among the three age groups children (%).

Variables		β-thalassemia patients	Healthy Control children	P value
		N (%)	N (%)	
Terminal plane	Mesial step	19(79.2)	21(87.5)	0.192
	Flush	2 (8.3)	3 (12.5)	
	Distal step	3 (12.5)	0 (0)	
Angel's Classification	Class I	24 (80)	25(83.3)	0.739
	Class II	6 (20)	5 (16.7)	
	Class III	0 (0)	0 (0)	
Horizontal overjet	Normal	27(50.9)	43(79.6)	0.004**
	Increased	20(37.7)	6 (11.1)	
	Decreased	6 (11.3)	5 (9.3)	
Vertical overbite	Normal	45(84.9)	48(88.9)	0.084
	Increased	2 (3.8)	5 (9.3)	
	Anterior open bite	6 (11.3)	1 (1.9)	
Spacing in maxilla	Normal	21(38.9)	23(42.6)	0.334
	Spaced	24(44.4)	27 (50)	
	Crowded	9 (16.7)	4 (7.4)	
Spacing in mandible	Normal	27 (50)	35(64.8)	0.101
	Spaced	14(25.9)	14(25.9)	
	Crowded	13(24.1)	5 (9.3)	

p value significant ≤0.05

Table 5 showed the distribution of malocclusion among the three age groups. Children with β-thalassemia major revealed various types of malocclusion. A statically significant correlation existed between β-thalassemia major and healthy control children in increased overjet.

Discussion

The relationship between thalassemia and malocclusion has been well documented in the literature [3,20-23].

In the present study children with β-thalassemia major exhibited various types of malocclusion; open bite (11.3%), increased overjet (37.7%), maxillary spacing (44.4%), maxillary crowding (16.7%), mandibular spacing (25.9%), and mandibular crowding (24.1%).

Angle's class I was the most frequent occlusal relation (80.0%) in children with β-thalassemia major, (20.0%) had class II occlusal relation, and no one had class III Angle's classification. In comparison with the healthy control group a slightly more percentage had Angle's class I occlusal relation (83%) as well no one had Angle's class III. However, no statically significant difference existed between the two study groups in relation to all types of malocclusion except for overjet (p≤0.004).

The results of occlusal relationship in this study was in agreement with the results by Elangovan et al., [5] in India among 72 children 6-18 years old with β-thalassemia major. However, a study carried by Gubta et al in India among 100 β-thalassemia major children 12-17 years old, showed higher percentage of Angle's class II (55% vs 15.7%) when compared to normal children, while Hattab et al reported class II skeletal base relation among all 54 Jordanian children with β-thalassemia major 5-16 years old [21,23].

A study carried out by Salem et al. [20] in Iran showed that a significant difference existed between 66 β-thalassemia patients and 66 healthy controls in overjet and open bite. Angle's classification of malocclusion was more or less the same percentage as in the present study class I (83.3%) and class II (19.7%). The percentages of various types of malocclusion were more than the percentages in the present study; open bite (12.1%), overjet (40.0%), maxillary crowding (30.3%), whereas the maxillary spacing (15.2%) was much less than that reported in the present study (44.4%).

Salem et al concluded that facial bones in patients with β-thalassemia major specially the maxillary bones undergo hyperplasia, to compensate for anaemia and hypoxia, which may be responsible for various degrees of malocclusion in β-thalassemia major patients [20].

In accordance to the present study Mehdizada et al. conducted a study among Iranian population aged 2-20 years old, different degrees of

malocclusion openbite, and deepbite were seen in thalassemia patients especially in the elder group 13-20 years old. This may be due to the early hypertransfusion treatment in young patients [22].

The results of the current study showed (37.7%) of the children with β-thalassemia major had increased overjet, which was consistent with Abu Alhaija et al. [23] results (37.8%) among 54 Jordanian children with β-thalasemia major aged 5-16 years old. In contrast Hattab FN reported less percentage of increased overjet (25.9%) among Jordanian children with β-thalassemia major [3].

Furthermore, Abu Alhaija et al reported increased overbite in (16.2%), anterior open bite in (21.6%) and normal overbite in (29.7%) individuals with β-thalassemia major [23]. Whereas, the present results showed less percentages of increased overbite (3.8%), anterior openbite (11.3%), and normal overbite (84.0%) in children with β-thalassemia major.

In the present study anterior openbite occurred more frequently among patients with β-thalassemia major than the control group. The same finding was noted in a study by Malath et al in Baghdad in which deepbite was more frequent among the β-thalassemia children in comparison to the healthy control group which was in contrast with the present findings [24].

The variation in the results among different populations may be attributed to the sample size, environmental factors, methods of assessment of malocclusion and age of the study population.

Limitations

- The number of children enrolled in the study was diminutive.
- Blood investigations should be carried for the control group children to ensure all of them are free of the disease and not just rely on the family history.

Conclusion

- Various types of malocclusion were perceived in β-thalassemia major children in Sudan; increased overjet and anterior open bite were the most frequent.
- A significant correlation existed between β-thalassemia major and healthy control group in increased overjet (p=0.004).
- The prevalence of malocclusion in β-thalassemia major increased with age.

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