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Original Articles

Prevalence of Orofacial Complications in Iranian Patients with β-Thalassemia Major

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Abstract

Background: Major β-thalassemia is the most common monogenic known disorder in Iran and about 15000 persons are affected. The purpose of this study was to evaluate the prevalence of orofacial complications.

Methods: In this cross-sectional study during 2003-2004, 300 patients with major β-thalassemia referring to 8 hospitals for routine examinations, blood infusion and treatment in cities of Tehran, Isfahan, Sari, Zahedan, Shahre Kord and Sanandag were studied. The questionnaires included general information, medical and dental history.

Results: The prevalence of orofacial complications in major β-thalassemia was: saddle nose (67%), maxillary protrusion (49/7%), color change of oral mucousa (41/7%), rodent face (34/7%), deep bite (21/7%), spacing (20/7%), and open bite (8/7%), respectively. There was not any relationship between the complications and sex. Most of the patients (91%) were in the first and second decade of life. Early diagnosis and blood infusion caused less prevalence of rodent face. The most observed type of occlusion was class 1, 2 and 3, respectively.

Conclusion: Knowing the prevalence of complication can help the dentists to do properly for these patients to solve their problems and improve knowledge of the parents about their children's dental health.

Keywords: *Major β- thalassemia, Orofacial complications, Iran*

Introduction

The thalassemias are a group of congenital disorders characterized by a deficient synthesis of either the α or β chains of globin in the hemoglobin molecule. As a result, the red blood cells are microcytic and hypochromic with an aberrant morphology. (1-2) The homozygous type that is known as β- thalassemia major or Cooley's anemia is the most common monogenic disorder in the Mediterranean basin, the Middle East, Asia and the south pacific. In Iran, about 15000 individuals are affected (3).

β- thalassemia major is the most severe congenital hemolytic anemia. At 4 to 6 mo of life, with the change from fetal xx chain to adult xx chain hemoglobin production, the first clinical manifestations appear. The hematocrit decreases to less than 20, the degree of anemia can reach a hemoglobin level of 2 to 3 g/dl, and the hemolysis is extensive, as is the iron over load (1-4).

Growth and development in children is slow. In adolescence, secondary sex characteristics are delayed. The skin color becomes ashen-gray due to the combination of pallor, jaundice, and hemosid erosis. Patient also presents cardiomegaly, hepatomegaly, and splenomegaly (5).

Bimaxillary protrusion and other occlusal abnormalities are frequent in thalassemia major cases. Dental and facial abnormalities include spacing of teeth, open bite, prominent malor bones, protrusion of maxilla and saddle nose.

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In addition, the pneumatization of the maxillary sinuses is delayed. Because of these skeletal changes, the upper lip is retracted, giving the person a “chipmunk face " or “rodent face"(6).
In β-thalassemia major, there is no correlation between the chronologic, skeletal and dental age. The skeletal retardation increases with age due to hypoxia from severe anemia, endocrine hypofunction secondary to iron deposition, or the toxic action of iron enzyme systems leading to tissue injury.
The oral mucosa is pale or lemon yellow color due to anemia and deposition of billirubin pigment then decrease lysis of red blood cells cause less deposition of billirubin (1-5).
The purpose of this study was to evaluate the prevalence of orofacial complications in Iranian Patients with β-Thalassemia major.

Materials and Methods

In this cross sectional study during years 2003-2004, 300 patient with β-thalassemia major (158 males and 142 females) in all ages referred to the special centers in 8 provinces of Iran for blood transfusion including (3 centers in Tehran, Sari, Isfahan, Zahedan, Sanandage, and Shahre kord) were participated. The centers were selected as cluster form different parts of Iran with a number of at least 35 patients in each province.
Provided questionnaires included all information (General, medical and dental). Clinical dent orofacial examination of the patients was performed by means of disposable mirror, probe, flash light and sterilized gauze.
Data were analyzed through the statistical tests of X2 (chi-square) considering $P< 0.05$.

Results

The number of patients with β-thalassemia major were 300, 158 (53/7%) males and 142 (47/3%) females from all age groups. Most of them (91%) were in the first and second decade of life.
There was not any relationship between the complications and sex. The prevalence of orofacial complications of β- thalassemia major patients has been shown in Table 1.
The most observed type of occlusion was class I, II, III, respectively (Table 2).
Relationship between prevalence of orofacial complications and thalassemia major patient's age of has been shown in Table 3 and 4.

Table 1: Prevalence of orofacial complications in patients with β - thalassemia major

Parameter number percent	Yes	No
	n (%)	n (%)
Rodent face	104 (34.7)	196 (65.3)
Saddle nose	201 (67)	99 (33)
Maxillary protrusion	149 (49.7)	151 (50.3)
Maxillary anterior teeth spacing	62 (20.7)	238 (79.3)
Anterior open bite	26 (8.7)	274 (91.3)
Deep bite	65 (21.7)	235 (78.3)
Mucousal discoloration	125 (41.7)	175 (58.3)

Table 2: Prevalence of occlusion classification in patients with β-thalassemia major

Classification of occlusion	Class I	Class II	Class III	Unknown
n	170	67	6	57
%	56.7	22.3	2	19

Table 3: Prevalence of orofacial complications in β-thalassemia major according to patient's age

Parameter \ Age (yr)	0-7	8-14	15-20	20<	*P*-value
	n (%)	n (%)	n (%)	n (%)	n (%)
Rodent face	11 (22.4)	32 (29.9)	46 (39.3)	15 (55.6)	0/014
Saddle nose	30 (61.2)	68 (63.6)	82 (70.1)	21 (77.8)	0/356
Maxillary protrusion	17 (34.7)	55 (51.4)	60 (51.3)	17 (63)	0/088
Maxillary Anterior teeth spacing	13 (26.5)	26 (24.3)	18 (15.4)	5 (18.5)	0/266
Anterior open bite	7 (24.3)	13 (12.1)	5 (4.3)	1 (3.7)	0/012
Deep bite	3 (6.1)	28 (26.2)	26 (22.2)	8 (29.6)	0/026
Mucousal discoloration	8 (16.7)	47 (43.9)	55 (47)	14 (51.9)	0/002

Table 4: prevalence of occlusion classification according to patient's age

Occlusion Number percent \ Age(year)	0-7	8-14	15-20	20<
Class I	23	70	65	13
	46.9	65.4	55.6	48.1
Class II	3	28	29	7
	6.2	26.2	24.8	25.9
Class III	0	0	3	3
			2.6	11.1
Unknown	23	9	20	4
	46.9	8.4	17.1	14.8

Discussion

In this study, most of the patients with β- thalassemia major were in the first or second decodes of life, which indicates a lack of life expectancy.

The study showed that saddle nose, protrusion, of maxilla and finaly "rodent face" or "chipmunk face" are the most common maxillo facial complications, respectively as stated in literature (7, 8).

In this research, most of the patients had class I occlusion (56.7%) which was compatible with other related studies in Iran and it was approximately similar to normal Iranian occlusion prevalence (70%) (9). The prevalence of class II was about 22.3% which was compatible with some other studies but prevalence of class II occlusion in normal population is about 17% so it can be concluded that class II occlusion prevalence in β- thalassemia major is more than the normal population (9, 10).

In this study, the prevalence of occlusion class III was 2%. In different reports stated percentage are 1%-6%. (10, 11). In normal population of Iran, this rate is about 6%, (15) which shows its abundancy in patients with major β- thalassemia is slightly less than normal population (9-11). Ninety percent of patients had unknown occlusion.

In clinical examination, a tint of lemon color was observed in oral mucosa due to existing billirubin produced by the decomposition of red cells. In

this study, deep bite was more than the normal population especially in older patients, which is due to rotation of mandibule and pro-trusion of maxilla with over growth of anterior teeth of maxilla. The anterior open bite was slightly more than normal population (12). Nevertheless, the difference was not significant.

The prevalence of orofacial complications in patients with β - thalassemia major had similarities and differences with other researches (13, 15). Overall, it indicated a reduction in complications during last decades, which was due to early diagnosis, treatment, and regular follows up.

Regular and repeated blood-infusion preserving the hemoglobin amount in an appropriate level (at least 10g/dl), along with iron removal can prevent face and skull deformities. Therefore skull and face deformities can be closely related to the patient's age, the intensity of anemia and the beginning time of treatment.

Conclusively, patients receiving inadequate blood transfusion in childhood will face more bone changes (expansion and deformity) in adolescence are cause of hyper activity of bone marrow to compensate anemia. Then early diagnosis and blood infusion cause less prevalence of complications.

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