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## **Original Article**

# Orodental complications and orofacial menifestation in children and adolescents with thalassaemia major of western Rajasthan population: a comparative study

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#### ARTICLEINFO

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#### ABSTRACT

AIM: This study was conducted to assess the prevalence of dental caries, malocclusion and facial changes in thalassemia major patients of western Rajasthan population. Methods: In this cross-sectional study, 50 patients with thalassemia major, and 50 healthy matched control individuals, were included. Dental caries indices, DMFT and DMFS were measured and compared with age matched healthy controls. Data were analyzed by t-student test. Results: Poor oral hygiene was generally observed. Dental caries were significantly higher in thalassemic patients (p<0.0001) in comparison with the healthy control group. Various malocclusion stages were seen. Facial changes were also seen (mongoloid features due to prominence of cheek bones, protrusion of the maxillary anterior teeth, and the depression of the bridge of the nose). Conclusion: Patients with  $\beta$ - thalassemia major showed highly significant differences in orodental and orofacial manifestations as compared to control group.

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#### 1. Introduction

The word 'Thalassa' in Greek means 'the sea' and 'Haema' in Greek means 'blood'. The word originated from a common condition peculiar to the geographical area in the Mediterranean between South Europe and North Africa.

Thalassemia was not recognized as a clinical entity until 1925, when Thomas Cooley, a Detroit pediatrician, described a syndrome among children of Italian descent characterized by profound anemia, splenomegaly and bony deformities [1].

-Thalassemia major or homozygous -thalassemia or cooley's anemia, the most severe form of -thalassemia is characterized by severe anemia and life-limiting complication in thalassemia major, however, no -chains are produced or only a very small amount is produced which prevents the synthesis of normal adult hemoglobin and severe damage the red blood cell's capacity to transport oxygen.The symptoms related to thalassemia include both physical signs as well as a variety of symptoms related to iron overload, change in hemoglobin concentration and dental problems. The oral structural changes occurs in thalassemic patients are due to maxillary enlargement resulted into protrusion of anterior teeth, increased space between teeth, overbite and open bite and varying degree of malocclusion which further predisposed to dental carries[2].

The orofacial manifestations of thalassemia are the result of bony changes occurring due to ineffective erythropoesis [3]. Overgrowth causes a characteristic appearance known as 'Chipmunk facies' [4].Dentofacial manifestations are a protrusive premaxilla associated with alveolar enlargement [5]. Beta Thalassemia Major is a hereditary hemolytic anemia disease with various grades of severity, which can be found with no or less globin chain qualitative synthesis. The patient often experiences hepatosplenomegaly, growth retardation and bone disorders and the Thalassemia facies / chipmunk face appearance. The orofacial manifestations of Beta Thalassemia major are prominent cheek bones and protrusive premaxilla due to erythroid hyperplasia with depressed bridge of the nose. The dentition shows protrusion, flaring and spacing of the maxillary anterior teeth, open bite that leads to malocclusion. The anemic condition makes the patient is difficult to do all oral hygiene instruction thus caries index will increase [6].Dental caries and orofacial changes in thalassemia major patients are reported by several investigators found that dental caries was significantly higher in thalassemia patients as compared to healthy controls [2,7,8,9,10].

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#### 2. Material and methods

The present study was carried out in the Department of Pediatrics,Umaid Hospital for women and children and Department of Anatomy, Dr. S.N. Medical College & Associated group of Hospitals, Jodhpur & also in Central Academy School Jodhpur.

Fifty (50) Healthy controls and fifty (50) thalassemic patients of both sex and varying age groups were included in the present study. These patients were also registered in Marwar thalassemia Society. These selected 50 thalassemic patients were regularly attending the Out Patient Department (OPD) or blood bank for repeated blood transfusion or admitted in the wards of Department of Pediatrics,Umaid Hospital for women and children, Regional Institute of Maternal & Child Health, Dr. Sampurnanand Medical College & Associated group of hospitals, Jodhpur.

The subjects selected for the study were grouped as:-A) Healthy controls (n=50) B) thalassemic patients (n=50) The subjects were further subdivided into following four groups: Group 1 Healthy control children. Group 2 Healthy control adolescents. Group 3 thalassemic children. Group 4 thalassemic adolescents.

All diagnosed thalassemic patients and Healthy controls were further evaluated for present study by taking Dental diagnosis The subjects selected for the study were confirmed for the following data:

- 1. They all were residents of western Rajasthan.
- 2. They had authentic documentation of their date of birth (i.e. birth certificate, hospital records)
- 3. Subjects from all strata of society were included

#### 2.1. General Examination

- 2.1. General Examination
  - The detailed information regarding:
- 1. Personal details of subjects (Name, Age, Sex, Religion etc.)
- 2. Family history (Carrier status of father, mother or siblings)
- 3. History of thalassemia (Age of diagnosis)
- 4. Dental Diagnosis (DMFT, DMFS scores).
- 5. Facial changes.
- 6. Blood groups & Hemoglobin (from patient's records and from student's record).
  - Each subject was examined for Dental diagnosis.

#### 2.2. Dental diagnosis :

1. Decayed-Missing-Filled Teeth Index (DMFT Index) DMFT Index was applied only to permanent teeth is composed of three components.

'D'- was used to describe decayed teeth.

'M'- was used to describe missing teeth due to caries.

'F'- was used to describe teeth that have been previously filled.

Mouth mirror and a fine-pointed pig-tail explorer were used for determine the sum of how many teeth were:

"Decayed," "Missing" or extracted due to decay, and filled as a result of caries involvement.

It was calculated by adding each component, i.e. D, M, & F.

#### 2.3. Decayed-Missing-Filled Tooth surfaces Index (DMFS)

Mouth mirror and a fine-pointed pig-tail explorer were used for determine the sum of how many tooth surfaces were: "Decayed," "Missing" or extracted due to decay, and filled as a result

The surfaces examined were:

of caries involvement.

- 1. For Posterior teeth: Five surfaces were examined and recorded: facial, lingual, mesial, distal and occlusal.
- 2. For Anterior teeth: Four surfaces were examined and recorded facial, lingual, mesial and distal. [figure.1]



Individual DMFS Index was calculated by: Total number of decayed surfaces = D

Total number of filled surfaces

DMFS score

Total number of de	cayeu sui laces	– D
Total number of mi	ssing surfaces	= M

- = F
  - =D+M+F [Soben Peter, 2003]

Arithmetic mean and standard deviations were calculated for both parameters studied. 'p'-values (probability) were determined to make out the statistical significance of variance between the mean values of individual parameters between the two groups of the subjects studied. p-value<0.0001, p-value<0.001, p-value<0.05 were considered the significant, highly and a very significant relationship respectively

#### 3. Result and Discussion

Certain medications may also interfere with vitamin D absorption or metabolism. Orlistat, a commonly used weight-loss drug and cholestyramine, a cholesterol lowering drug can reduce the absorption of vitamin D and other fat-soluble vitamins [59,60] Corticosteroids and epilepsy drugs Phenobarbital and dilantin interfere with the metabolism of vitamin d, reducing active levels [61,62].

Malocclusion was generally seen in the patients, especially in the older individual. The oral manifestations in thalassemia major were studied in present study; it was observed that commonest manifestation was pallor of mucosa. Malocclusion was also reported which was due to proliferation of marrow within the frontal and facial bones. Delayed eruption and dental caries were also reported in that study. High rates of malocclusion were reported in thalassemic patients [9, 11] but by early hypertransfusion treatment policy, it has been controlled in younger patients. In the present study out of 50 thalassemic patients 38 thalassemic patients had thalassemia facies / chipmunk face appearance. The orofacial manifestations of Beta thalassemia major are prominent cheek bones and protrusive premaxilla [7].

DMFT of thalassemic patients  $(3.3 \pm 3.03)$  shows highly significant difference (p-value<0.0001) when compared to the DMFT of healthy controls  $(0.52 \pm 0.88)$  [table.1].DMFT of thalassemic children  $(3.13 \pm 3.21)$  and thalassemic adolescents  $(3.83 \pm 2.40)$  shows highly significant difference (p-value<0.0001) when compared to the DMFT of healthy control children  $(0.48 \pm 0.96)$  and healthy control adolescents  $(0.58 \pm 0.77)$  [table.3]

TABLE NO.1 shows mean DMFT in the two groups

Group studied	DMFT Score	Statistical analysis	
	Mean ± S.D [Range]	p-value	Statistical significance
Healthy controls (50)	0.52±0.88(0-2)		
Thalassemic patients (50)	3.3±3.03(0-12)	<0.0001	HS

HS-Highly Significant

Similarly, Mehdizadeh performed their study on 50 patients (21 male and 29 female) with thalassemia major between 2and 20 year of age and 50 healthy control individual were included. They studied the orodental complications in -thalassemia; it was found that oral hygiene condition evaluated by plaque index was not good. Dental caries were significantly higher in thalassemic patients (p<0.001) in comparison with healthy control group. It was concluded that thalassemia is associated with higher rates of dental carries and malocclusion [10]. Also Hattab proposed their study on 54 thalassemic patients. The prevalence of dental caries in the thalassemic patients was considerably higher (22.7%) than that reported in a normal Jordanian sample (DMFT 6.26 vs. 4.84). it was also observed that more than half of patients had poor oral hygiene [9].

A highly significant difference (p-value<0.0001) is observed in the DMFS of thalassemic patients ( $6.22 \pm 6.79$ ) and Healthy control subjects ( $1.28 \pm 2.03$ ) [table.2]. DMFS of thalassemic children ( $6.42 \pm 7.54$ ) and thalassemic adolescents ( $5.58 \pm 3.73$ ) shows a very significant difference (p-value<0.001) when compared to the DMFS of healthy control children ( $1.06 \pm 1.98$ ) and healthy control adolescents ( $1.63 \pm 2.11$ ) [table.4]. Similarly Al-Jobouri conducted their study on 41 Beta thalassemia major patients (31 male, 10 female) and 41 healthy control subjects. This study showed that Beta thalassemia group had higher DMFS. The DMFS score for thalassemic major group was  $9.29 \pm 7.66$  and for control group DMFS was  $2.54 \pm 2.44$ . The thalassemia major group had higher

### TABLE NO.2 shows mean DMFS in the two groups

Group studied(n)	DMFT Score	Statistical analysis	
	Mean ± S.D [Range]	p-value	Statistical significance
Healthy controls (50)	1.28±2.03(0-6)	0.0004	
Thalassemic patients (50)	6.22±6.79(0-24)	<0.0001	HS

HS-Highly Significant

#### ABLE NO. 3 shows mean DMFT in the four groups

Group studied	Healthy controls Mean ± S.D	Thalassemic Patients Mean ± S.D	p-value	Statistical analysis Statistical Significances
Children (0-10 yrs)	$0.48 \pm 0.96$	3.13±3.21	< 0.0001	HS
Adolescents (11-17 yrs)	0.58±0.77	$3.83 \pm 2.40$	<0.0001	HS

HS-Highly Significant

#### TABLE NO. 4 shows mean DMFS in the four groups

Group studied	Healthy controls Mean ± S.D	Thalassemic Patients Mean ± S.D	p-value	Statistical analysis Statistical Significances
Children (0-10 yrs)	$0.48 \pm 0.96$	$6.42 \pm 7.54$	< 0.0001	VS
Adolescents (11-17 yrs)	1.63±2.11	$5.58 \pm 3.73$	<0.001	VS

VS-very significant

#### 4.Conclusion

DMFT and DMFS indices were significantly higher in thalassemia group, especially in older patients. This could be due to poor oral hygiene, poor motivation, endocrine problems, and immune deficiency. A low IgA level in saliva has also been shown in thalassemic subjects, which may contribute to an increased rate of dental caries and prevalence of orofacial changes could be figured out simultaneously

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