ORIGINAL ARTICLE

Oro-facial Manifestations, Oral Health Status and Treatment Needs in Transfusion Dependent Children With β-thalassemia Major: A Cross-sectional Study

Ankita Arora[1], Savitha Sathyaprasad[2]

[1] Department of Pediatric dentistry, Faculty of Dentistry, Manipal University College Malaysia, Jalan Batu Hampar, Bukit Baru, 75150, Melaka, Malaysia.
[2] Department of Pedodontics and Preventive Dentistry, KVG Dental College and Hospital, 574239, Sullia DK, India

ABSTRACT

Introduction: β-Thalassemia is genetic disorder which clinically presents as anaemia due to decreased production of beta-chains of haemoglobin molecule. Literature on oro-facial manifestations, oral health status is sparse and inconclusive in the β-thalassemia major patients (TM). **Methods:** This cross-sectional study was done to assess oro-facial manifestations of β-Thalassemia. 31 TM patients in the age group of 6-18 years were clinically examined and oro-facial manifestations were recorded. Oral health status including oral hygiene status, dental caries, gingival status of these TM children was compared with 42 healthy children. The mean values were compared using Student's t-test. Treatment needs of both the groups were recorded using WHO oral health assessment form. **Results:** Lip incompetence (38.75%) and maxillary protrusion (19.35%) were most common oro-facial manifestations of Thalassemia major. Mean OHI-S score for TM and healthy children were comparable 1.73 ± 1.10 and 1.36 ±1.17 respectively (p=.178) but mean GI score for TM was higher than healthy children, 1.42±.39 and .94±.51 respectively (p<.05). Mean DMFT scores for TM and healthy children were 1.5±1.8 and 1.2±2.1 respectively (p=.578). Treatment needs was relatively higher among TM children with 58% of these children requiring one surface filling. **Conclusion:** Lip incompetence followed by maxillary protrusion were most common oro-facial manifestations of Thalassemia major. There is no significant association between oral hygiene and dental caries with Thalassemia major. However, thalassemia major is associated with gingivitis. Treatment requirement was relatively higher among TM children with one surface filling being the commonest need.

Keywords: Thalassemia Major, Children, Orofacial Manifestations, Oral health status, Treatment Needs.

Corresponding Author:
Ankita Arora, MDS
Email: ankita.arora@manipal.edu.my, 4ankitaarora@gmail.com
Tel: +60172596982

INTRODUCTION

Thalassemia are a diverse group of genetic blood diseases caused by defects in the synthesis of one or more of the globin protein chains of the haemoglobin molecule with extensive haemolysis resulting in microcytic anaemia of varying degrees with a haemoglobin level of 2 to 3 gm/dl (1-3). The disorders are classified either based on defective globin chains (α or β) or based on the severity of clinical signs and degree of anaemia (thalassemia major, thalassemia intermedia, thalassemia minor) (1-3). The prevalence rate of Thalassemia is 4% in Asian Indians (4). Beta – thalassemia is caused by point mutations or by deletions in the beta globin gene on chromosome 11, leading to the reduced or absent synthesis of beta chains of haemoglobin (1,2). The homozygous form of β-thalassemia is also known as Cooley's anemia or Mediterranean anemia (1-5). Clinical presentation includes pallor of the skin and mucosa, growth retardation, a high risk for infection and hepatosplenomegaly (1-3, 6). Bone marrow transplantation is the only definitive cure currently available for thalassemia which has a multitude of problems like finding a matching donor and variable success rates (1-3, 6). Other medical treatments that help in regulating low haemoglobin levels are regular blood transfusions, iron chelation, and management of secondary complications of iron overload.

Pallor of the mucosa, saddle nose, maxillary protrusion, rodent face and malocclusion caused by expanding marrow spaces are some of the reported orofacial complications of this disease (4-6). Thalassemia may have a profound effect on the health of the oral tissues. The aim of this study was to evaluate different oro-facial manifestations, oral health status and treatment needs of

a group of Indian children with β-TM and to compare that with healthy (thalassemia – free) children stratified as per age.

MATERIALS AND METHODS

This was a comparative, cross-sectional study. Ethical clearancewas obtained from the institutional ethical committee of K.V.G Dental College and Hospital, Sullia (Ref. no. MD/Pedo/O16/2011-12) and the National Governmental Organization – Foundation Against Thalassaemia (Regd), Faridabad, India.

A total of thirty-one (n=31) β-Thalassemia major patients in the age group of 6 to 18 years, registered with National Governmental Organization – Foundation Against Thalassaemia (Regd), Faridabad, India formed the study group. Forty-two healthy (n=42) children who met inclusion criterion were chosen from relatives and/ or friends of 'children with thalassemia' to match age, sex, and socioeconomic status.

Inclusion criterias for Group 1 includes β-thalassemia major patients, age between 6 and 18 years and obtained consent from parents. For Group 2, the includsion criterias include healthy children (age between 6 and 18 years) and consent from parents. The exclusion criteria were having dental prophylaxis in the last 6 months, presently on dental treatment (to eliminate bias in scoring of indices) and having comorbid conditions known to influence dental caries or the severity of periodontal diseases like diabetes.

Clinical examination and indices

All children were examined in a dental clinic within premises of a multi-specialty hospital. Following complete medical and dental history, all children were examined for oro-facial manifestations, dentition status and treatment needs, and gingival status while seated on a dental chair by a single trained and calibrated investigator. Prior to the study intra-examiner reliability was determined using Cohen's kappa statistics. k>.93 was used as an acceptable level in diagnosis of dental caries, OHI-s , GI-s and treatment needs. Recording of data was done by a trained dentist who assisted throughout the study. The clinical examiner recorded oral manifestations (if any present). Clinical assessment of dental caries was done visually using criteria recommended by the World Health Organisation and documented using the DMFT /deft index), Oral hygiene as per Oral Hygiene Index – Simplified (OHI-S) developed by Greene and Vermillion (8), Gingival status as per Gingival index (GI) developed by Loe and Silness (1963) (9,10). Treatment needs were assessed as per WHO Oral health assessment form (1997) (7).

The examination was done in a uniform manner beginning from the maxillary right quadrant in a clockwise direction in the maxillary and mandibular region. Oral hygiene and gingival scores were recorded prior to examination for caries.

Statistical analysis

The data obtained were entered using statistical package for social sciences (SPSS) version 20. The mean values were compared using Student's t-test. A p value of <.05 was considered significant. Prevalence of dento-facial anomalies (%) was determined for children with thalassemia. Descriptive details of treatment needs of both the groups are presented in tabular manner.

RESULTS

Table I describes the population distribution of children with Thalassemia and healthy children, total sample was comprised of 24 females (32.9%) and 49 males (57.5%). Oro-facial manifestations of the disease are presented in Table II. Lip incompetence (38.75%) followed by maxillary protrusion (19.35%) were most common manifestations in this study sample. Statistically significant differences in gingival index are reported in children with thalassemia when compared to healthy children when mean GI scores 1.42±.39 and .94±.51 respectively were compared (p<.05), this is despite non-significant difference of oral-hygiene index scores 1.73 ± 1.10 and 1.36 ±1.17 respectively of both groups (p=.178) as demonstrated in Table III.

Table I: The descriptive statistics of the sample population distribution

Variable	n (%)
Age (Mean±SD)	10.6±3.3
Gender	
Female	24 (32.9)
Male	49 (67.12)
Children with Thalassemia	31(42.55)
Healthy children	42(57.5)

Table II: Orofacial manifestations in children with thalassemia

Manifestation	n (%)
Saddle nose	6 (19.35%)
Lip incompetence	12 (38.75%)
Frontal bossing	4 (12.90%)
Maxillary protrusion	8 (25.80%)
"Chipmunk" facies	4 (12.90%)

Table III: Comparison of OHI-s, GI and DMFT scores between TM and Healthy children

	Children with Thalassemia Mean (SD)	Healthy Children Mean (SD)	Mean difference (95% CI)	t (df)	P value[a]
OHI-s	1.73 (1.10)	1.36 (1.17)	0.37 (-0.17, 0.91)	1.370 (71)	0.175
GI	1.42 (0.39)	0.94 (0.51)	0.48 (0.26, 0.70)	4.377 (71)	<0.001
DMFT	1.50 (1.80)	1.20 (2.10)	0.30 (-0.63, 1.23)	0.640 (71)	0.524

95% CI: 95% confidence interval; [a]Unpaired t-test
Statistical test applied: Student's t-test was used to compare Mean scores of the two groups.

In the current study, there were no significant difference in mean DMFT scores between TM and healthy children (P=0.578) as depicted in Table III.

Treatment needs of both the groups are presented in Table IV. Treatment needs was relatively higher among TM children than healthy children, with requirement of one surface filling being the commonest treatment need in thalassemia patients (42.85%).

DISCUSSION

The literature on orofacial manifestations in children with thalassemia is sparse and inconsistent. The association of thalassemia with oral conditions such as oral mucosal changes, gingival and periodontal diseases, and dental caries is debatable. Literature describes typical facial appearance of children with thalassemia as "chipmunk" like appearance or "rodent facies" with frontal bossing, saddle nose, high and bulging cheek bones, retraction of the upper lip and maxillary protrusion and spacing of other teeth (5,6,11-14). Proliferation of the bone marrow in the facial skeleton leading to the expansion of the facial bones, resulting in dental malocclusions and protrusions is the explained pathophysiology (11-14). The proliferated marrow acts as an ancil¬lary hematopoietic organ to compensate for the chronic haemolysis in these patients (5,6,11-14). The prevalence and severity of these atypical features in TM patients examined in our study are tabulated in Table II. Most cases had normal facies which could be attributed to regular blood transfusions in them. As the cases reported regularly for blood transfusions and concerned medical treatment, maintenance of haemoglobin levels in normal clinical range supported normal growth and prevented the occurrence of craniofacial defects and lead to the development of normal facies. It is in accordance with findings of a study by Cutando A and Jirarattanasopa V et al that reported that adequately transfused subjects did not express typical thalassaemic facies or appearance and had class I molar and incisor relationship with normal overjet and overbite while patients transfused occasionally showed craniofacial abnormalities and malocclusion (6, 15).

Pallor of mucosa, glossodynia and loss of papillae on the tongue are some of the reported oral manifestations (16-18). However, our study did not show any significant above mentioned oral manifestations of thalassemia major in its patients which could be attributed to well-maintained haemoglobin levels in the study group (Table II). Mattia D et al also reported the prevalence of pallor of oral mucosa to be as low as three out of sixty TM cases examined, which is similar to our study (14). In our study, oral hygiene levels in thalassemia patients (Table III) could be attributed to negative attitude and poor dental knowledge of thalassemic subjects and their parents toward proper oral hygiene and dental health. These results are similar to findings of studies by P. F. Luglиn et al and Al-Wahadni et al who reported the difference in oral hygiene of cases and controls as not statistically significant (19,20).

In our study mean gingival score difference between the two groups was statistically significant. ($p < 0.001$)(Table III). The explanation for increased gingival inflammation in thalassemia group can be attributed to iron overload in these patients owing to repeated blood transfusions. Caliskan U et al reported iron accumulation in gingival tissues of thalassemia patients and correlated them to their poor periodontal status (21). Furthermore, a literature review on virulence factors of P. gingivalis, a periodontopathogen and an established agent in producing gingival bleeding describes its virulence mechanism owing to the production of haemagglutinin, hemolysin and several enzymes by these bacteria that may promote colonization by aiding in the acquisition of hemin and Iron (22). Further research on gingival iron levels, variety, and intensity of colonization in oral tissues in thalassemic patients might explain the theoretical risk of gingival diseases in these patients. The findings of our study regarding prevalence and severity of gingival diseases in thalassemia patients are similar to the findings of study done by Hattab FN (13) but contrast with a study by Al-Wahadni et al (20) where in results showed no significant differences in plaque index and gingival index between thalassemia patients and healthy control groups concluding that Thalassemia is not associated with increased rates of gingivitis or

Table IV: Treatment needs in Children with Thalassemia and Healthy children

Treatment Needs	Children with Thalassemia		Healthy children	
	No. of persons N (%)	No. of teeth	No. of persons N (%)	No. of teeth
One surface filling	18(58)	33	8(19.04)	21
Two or more surface filling	6(19.3)	11	5(11.90)	9
Crown for any reason	8(25.80)	22	7(16.66)	10
Pulp care and Restoration	8(25.80)	22	7(16.66)	10
Extraction	5(16.12)	6	3(7.14)	4
Fissure sealant and need for any Other care	10(32.23)	33	6(14.2)	20
Total(*)	55		36	

As single patient requires multiple treatments, the total number exceeds more than 42.

periodontitis.

Our study found no difference in caries prevalence (Table III) between thalassemia patients and healthy controls. This finding in our study was similar to findings of the study by Raju P et al (17). However, in contrast to our findings, Al-Wahadni AM et al (20), Gomber S and Dewan P (4) reported that thalassemia is associated with higher rates of dental caries. Luglie PF et al (19) found increased presence of mutans streptococci at detectable levels in TM patients.

To the best of our literature search, this study was the first systematic attempt to identify treatment needs of these group of children. Treatment needs of TM patients were higher than healthy children where a maximum number of cases required one surface filling and about one-third of children require pit n fissure sealant and other preventive procedures like preventive resin restoration etc (Table IV). This finding suggests that most of the children with thalassemia had caries on one surface which could have been treated with timely intervention. Documentation done through our study regarding treatment needs of children suffering from Thalassemia major will help planning further treatment protocols to meet special oral health care concerns of this patient population. Preventive care like pit and fissure sealants, preventive resin restorations emphasized in the early stage can reduce morbidity as far as oral health is concerned. Alternative restorative treatment using materials such as glass ionomers that release fluoride may be an added benefit as both preventive and therapeutic approach.

CONCLUSION

Lip incompetence followed by maxillary protrusion were most common oro-facial manifestations of Thalassemia major. There is no significant association between oral hygiene and dental caries with Thalassemia major. However, thalassemia major is associated with gingivitis. Treatment requirement was relatively higher among TM children with one surface filling being the commonest need.

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