Original Article

Implications of β-thalassemia on oral health status in patients: A cross-sectional study

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Abstract

Background: β-Thalassemia is a chronic disease of autosomal recessive origin that is identified by the presence of a severe form of anemia. This hematological disease has been shown to directly influence a person's physical as well as psychological well-being along with their families. **Aim:** This study aimed to find an association between dental health status and oral health-related quality of life (QoL) among children who have been diagnosed with β-thalassemia. **Materials and Methods:** This prospective cross-sectional study was carried out in the dental outpatient department; blood bank and pediatric outpatient departments that were associated with the primary institute. All study participants were age-ranged from 3 to 15 years. Informed written consent was obtained from caregivers or parents of all the study participants. This study was conducted for a total duration of 1 year (from June 2020 to June 2021). All study participants were categorized into two groups: (a) Group I (n = 150) comprised children who were diagnosed with β-thalassemia and (b) Group II (n = 150) comprised normal controls. Exclusion criteria in the study included children suffering from any systemic disease that predisposes them to dental caries or periodontal diseases. The intra-oral examination was performed using Decayed-Missing-Filled Teeth Index (DMFT/dmft Index) and Oral Hygiene Index-Simplified (OHI-S). Assessment of QoL was done by using the "Child Perceptions Questionnaire for children." Collected data were recorded in Microsoft Excel workbook, 2007. Statistical comparison between both the groups was performed by using statistical tools such as the Chi-square test, Fisher's exact test, independent t-test, and Mann–Whitney U test. The probability values lesser than 0.05 were considered to be statistically significant. **Results:** Maxillofacial findings—rodent facies, saddle nose, lip incompetence, pale oral mucosa, anterior open-bite, lower anterior teeth crowding, and maxillary anterior teeth spacing or crowding—were seen. Class II malocclusion was present in significant numbers of subjects. On comparing dmft/DMFT scores, no significance was observed while on comparing OHI-S index, statistical significance was seen. A statistically significant difference in the QoL was noted between thalassemic children and the control group. **Conclusion:** Thalassemic children showed a significant association between dental health and QoL.

Keywords: Dental health, oral health status, quality of life, thalassemia

Introduction

Hemoglobinopathies which comprise thalassemia along with sickle-cell anemia have been reported to have widespread. Approximately 5% of the global population is found to possess genes that are responsible for various hemoglobinopathies.[1]

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Thalassemia is an autosomal recessive blood disorder that was first recognized in 1925 by Dr. Thomas Cooley.[2] This defect causes alteration in hemoglobin synthesis manifesting in variable degrees of anemia that ranges from clinical alterations those threatening to life. This hereditary type of anemia is the result of mutations in chromosome 16 that encodes globin genes and chromosome 11 encoding ϒ, δ, and β-globin. This particular defect in the globin chain may affect either α- or β-polypeptide chain of the globin portion of hemoglobin.[3]

Based upon the affected globin chain, thalassemia may be classified into α- or β-thalassemia. It may be also be classified into homozygous, heterozygous, or compound heterozygous based upon clinical and genetic defects.[3]

The heterozygous form of β-thalassemia disease/thalassemia minor is a mild form with minimal clinical manifestations while the homozygous form of β-thalassemia or thalassemia major demonstrates a severe form of clinical manifestations associated with distinct oro-facial defects. Under rare circumstances, a less severe form of the disease called "thalassemia intermedia" has also been reported. There are approximately 240 million genetic carriers of β-thalassemia all over the world. In India, the total number of patients with β-thalassemia ranges from about 30 million with a mean prevalence of 3.3%. The initial clinical symptom is seen at 4–6 months of life. The affected infants suffer from a severe form of anemia, show failure to thrive, and show progressive paleness. Other health issues include problems in feeding, diarrhea, the recurrent onset of fever, spontaneous bone fractures, bleeding, susceptibility to infections, hepato-splenomegaly, and growth retardation.[3]

Thalassemia causes a reduction in the rate of synthesis of anyone globin chains that is responsible for the synthesis of defective hemoglobin which results in anemia. As a result of this defect in globin chain synthesis, there is either no hemoglobin chains production or minimal amount gets produced which causes prevention of the synthesis of normal adult hemoglobin that leads to severe destruction of red blood corpuscles, thus reducing the ability for transportation of oxygen. The changes in the maxillofacial skeleton and dental tissues in thalassemic patients are mainly due to the result of enlargement of the maxilla that causes maxillary protrusion of anterior teeth, increase in spacing between teeth, and development of over-bite along with open bite, thus resulting in variable degrees of malocclusion that predisposes an individual to the development of dental caries.[4] The occurrence of dental caries in thalassemic patients may be attributed to improper diet, presence of malocclusion, lack of adequate knowledge regarding oral and dental health, reduction in the salivary concentration of urea, as well as levels of salivary immunoglobulin A (IgA).[5]

The most common oro-facial manifestation in β-thalassemia major (BTM) includes prominent frontal bossing and zygomatic arches, overgrowth of the maxillary bone, dental, and skeletal malocclusion along with delay in the development of teeth.[6]

Major changes seen in the facial region in thalassemia patients include the prominence of zygomatic bones and maxillary enlargement due to erythroid hyperplasia that is accompanied by depression of the nasal bridge. All of these changes may cause characteristic facial appearance referred to as "Chipmunk or Rodent facies." Other dental as well as facial defects include spacing between the teeth, forward drifting of maxillary incisors, anteriorly located open bite, protruding maxilla, malocclusions, and saddled nose. Also, pneumatization of the maxillary sinus is shown to exhibit a delay in affected subjects.[3]

β-Thalassemia patients are at a higher risk of developing dental caries as well as periodontal diseases.[7] This increased prevalence is explainable on the basis of the chronicity of this disease as the patients remain pre-occupied with this life-threatening disease and, as a result, neglect their basic and preventive dental health care. Other causes include inadequate knowledge of oral health, improper diet, and malocclusion.[8,9]

β-Thalassemia is the most commonly found variety of thalassemia that affects approximately 60–80 million individuals in the world, and approximately 3% of the total population around the world is carrying the β-thalassemia gene.[10]

Thus, based upon different oro-facial changes along with dental diseases, this study was designed to find out any association between dental health status and oral health-related quality of life (QoL) among children diagnosed with β-thalassemia.

Materials and Methods

This cross-sectional study was conducted in the dental out-patient department, blood bank, and pediatric outpatient department associated with the institute. The study participants were aged between 3 and 15 years. Informed written consent was obtained from the caregivers or parents of all the patients. The study was conducted for a duration of 1 year (June 2020 to June 2021). The study subjects were categorized into two groups: (a) Group I (n = 150) comprised children diagnosed with β-thalassemia and (b) Group II (n = 150) comprised control subjects.

Ethical approval for conducting this study was obtained from appropriate Research and Ethics Committee of the institute (IEC/22/21)

Exclusion criteria for the study were patients who suffered from any other disease that can result in dental caries or affect the severeness of periodontal diseases.

A thorough general physical examination was performed that included demographic data and a complete intra-oral examination. Oral health and dental health status were examined using the following indices:

(1) Decayed-Missing-Filled Teeth Index (DMFT/dmft Index), for both permanent and primary teeth.

(2) Oral Hygiene Index-Simplified (OHI-S).

Mouth mirror and curved explorer were used for the examination of teeth and oral cavity by a single operator throughout the study period. Data collection was performed by the intra-oral examination and interviews for assessment of the QoL using the "Child Perceptions Questionnaire for children."

All collected data were entered in Microsoft Excel worksheet 2007. Statistical comparison between both the groups was performed using statistical tools such as Chi-square test, Fisher's exact test, independent *t*-test, and Mann–Whitney's *U* test wherever was considered appropriate. The confidence interval was set at 95% while the accepted margin of error was set at 5%. The probability values lesser than 0.05 were considered significant.

Results

(a) On analyzing the prevalence of various dental and facial findings in the study, the following observations were made: rodent facies was evident in 49.1% males and 34.3% females with thalassemia. Saddle nose was evident in 64.6% and 54.1% male and female thalassemic patients; lip incompetence was noted in 91.5% and 70.5% males and females, respectively; pale-colored oral mucosa was notable in 22.1% and 24.4% male and female thalassemic subjects; the anterior open bite was seen 25.2% and 13.2% male and female patients; lower anterior teeth crowding was observed in 13.2% and 9.3% male and female thalassemic individuals while maxillary anterior teeth spacing or crowding was seen in 13.2% and 2.1% male and female thalassemic patients, respectively. Also, those children diagnosed with β-thalassemia showed a greater percentage of class II malocclusion (35.9%) when it was compared with the control subject group children (10.0%). A statistically significant difference ($P = 0.03$) was observed in the comparison between both the groups [Table 1 and Graph 1].

(b) On observing the OHI-S, dmft, and DMFT indices in thalassemic patients following mean ± standard deviation (SD) values were obtained: (i) mean OHI-S scores for males and female subjects were 2.68 ± 1.51 and 2.45 ± 0.84, respectively. Mean deft scores for male and female thalassemic subjects were 5.76 ± 4.01 and 5.96 ± 2.32, respectively, while mean DMFT scores for male and female thalassemic patients were 8.57 ± 2.08 and 5.13 ± 2.31, respectively. On inter-group comparison, a *P* value of 0.05 (statistically significant) was observed in the OHI-S index; however, on comparing dmft values, no significance ($P = 0.06$) was noted. Although on comparing mean ± SD values of DMFT scores, a statistically significant *P* value (0.01) was obtained [Table 2].

(c) On analyzing the QoL in these thalassemic children, a statistically significant difference ($P = 0.02$) was noted between thalassemic children and control group subjects while oral symptoms' analysis showed no significant difference ($P = 0.06$), functional limitations were demonstrated to have statistical significance ($P = 0.01$), emotional well-being showed no significant difference ($P = 0.07$), and social well-being was statistically significant ($P = 0.02$) [Table 3].

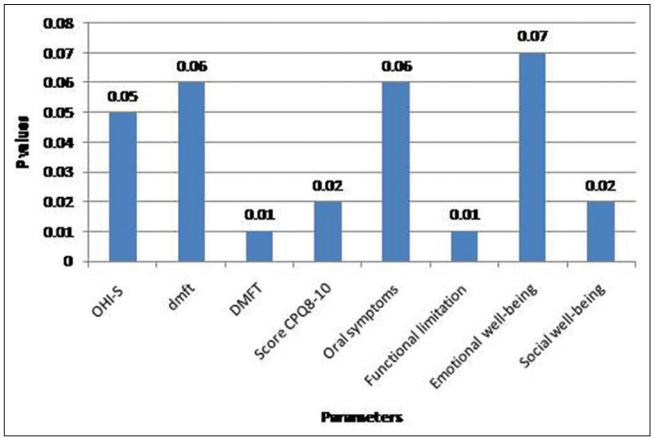
Table 1: Prevalence of dental and facial findings in the study

Parameter	Male *n* (%)	Female *n* (%)
Rodent face	20 (49.1)	12 (34.3)
Saddle nose	30 (64.6)	15 (54.1)
Lip incompetence	39 (91.5)	24 (70.5)
Pale oral mucosa	10 (22.1)	9 (24.4)
Anterior open bite	11 (25.2)	6 (13.2)
Deep bite	14 (27.1)	10 (23.1)
Lower anterior Teeth crowding	6 (13.2)	4 (9.3)
Maxillary anterior Teeth spacing/crowding	7 (13.2)	2 (2.1)
Class II malocclusion	18 (32.9%)	5 (10%)

Discussion

The term "Thalassemia" has been derived from the Greek word "thlassa" which means sea and "hemia" which denotes "blood." This term was first used by Wipple and Bradford (1932).[11] BTM afflicted patients report the severest form of clinical symptoms and include significant oral and facial defects.[12] The risk of developing various oral and dental diseases in thalassemic patients is very high. Thus, preventive measures must be taken against these oral diseases and are therefore considered very important as it increases an individual's well-being and also helps in higher life expectancy among these patients. Also, the role played by sound oral health status has demonstrated an increase in QoL. Thus, a unified approach toward standard dental care is considered essential that includes close association among hematologist, pediatricians, and a dentist.

The dentist, especially a specialist pediatric dentist, plays a crucial role in educating thalassemic patients and their parents or caregivers with regards to preventing dental caries and regarding the importance of maintenance of good and adequate oral hygiene status.



Graph 1: P-values in the study

Table 2: Mean oral hygiene index-simplified (OHI-S) and caries scores of primary and permanent dentition

	Mean±SD scores (OHI-S) (thalassemia patients)	Mean±SD scores (OHI-S) (control subjects)	Mean±SD DMFT scores (thalassemia patients)	Mean±SD DMFT scores (control subjects)	Mean±SD DMFT scores (thalassemia patients)	Mean±SD DMFT (control subjects)
Male	2.68±1.51	1.34±0.2	5.76±4.01	4.32±1.2	8.57±2.08	3.23±1.2
Female	2.45±0.84	1.23±0.3	5.96±2.32	4.21±1.1	5.13±2.31	2.32±1.3
P	0.05		0.06		0.01	

Table 3: Child perception questionnaire subscale

	Maximum score	Children with β-thalassemia (*n*=150) Mean±SD	Children in control group (*n*=150) Mean±SD	*P*
Score CPQ8-10	100	13.2 (12)	18.8 (16)	0.02
Oral symptoms	20	6.7 (4.3)	5.7 (3.5)	0.06
Functional limitation	20	2.3 (2.3)	4.6 (3.2)	0.01
Emotional well-being	20	3.1 (4.3)	4.1 (4.9)	0.07
Social well-being	40	2.6 (3.2)	4.8 (6.1)	0.02

Our study results have shown that the children suffering from β-thalassemia have a higher percentage of class II malocclusion (35.9%) when compared to the control group (10.0%). A statistically significant difference was observed in comparing both the groups. Similar observations have been documented by Gupta *et al*.,[10] Sakshi *et al*.,[13] and Mehdizadeh *et al*.[14] who made use of Angle's classification for evaluating the prevalence of malocclusion among thalassemic patients. This finding can be attributed due to the hyperplasia of bone marrow that can occur due to chronicity of anemia which results in prominence of the maxilla and lack of pneumatization within the maxillary sinuses. Also, retrusion of the mandible may occur as a result of generalized retardation of growth in children with thalassemia.[11,12]

Dhote *et al*.[15] in their study reported significantly greater dental caries experience in thalassemic patients, also an increase in the prevalence of gingivitis along with the accumulation of plaque in patients with thalassemia major when compared to controls.

Elangovan *et al*.[3] in their study reported 59.7% of thalassemic patients with class I malocclusion, 23.6% showed class II variety of malocclusion, while none of the patients had class III type of malocclusion.

Fadel *et al*.[16] assessed the oral health status of children with BTM and their oral health-related quality of life (OHRQoL) in relation to the serum ferritin level. It was found that the children with BTM generally had high dental caries experience and gingival inflammation, yet an acceptable OHRQoL.

Khan *et al*.[17] conducted a study to investigate the dental and oral health status of Egyptian children with BTM and its impact on their quality of life (OHRQoL) in comparison to their normal counterparts. It was observed that the thalassemic children had a worse dental status than controls that had a negative impact on the emotional well-being aspect of the adolescent thalassemia group, but there was no negative impact of thalassemia itself as a disease on OHRQoL.

Ebeid *et al*.[18] assessed the oral health status of adolescents with β-thalassemia and its impact on their oral health-related QOL (OHRQoL). It was concluded that adolescent with β-thalassemia had high prevalence of class II malocclusion, dental caries, and poor oral hygiene level that resulted in negative impact on their emotional well-being aspect.

Kaur *et al*.[2] observed a higher incidence of dental caries in patients diagnosed with thalassemia when compared with normal children. However, no significant increase in the levels of gingival inflammation or gingivitis or accumulation of plaque was observed in β-thalassemia patients compared to the control subjects.

Similar findings have also been reported in thalassemic individuals belonging to the Iranian populace by investigators Mottalebnejad *et al*.,[19] Babaei *et al*.,[20] Ajami *et al*.,[21] and Shooriabi *et al*.[22] This high prevalence of dental caries can be attributed to the long duration of these hemoglobinopathies that are life-threatening as a result of which oral health care and dental hygiene may not be a major cause of concern among these patients.

Conclusion

In conclusion of this study, an insight into various oral as well as dentofacial manifestations of β-thalassemia has been evaluated. This study has reinforced the associations that exist between oral health status and various maxillofacial presentations along with problems in general systemic health among these patients. Thus, our study findings stress the importance of periodic and detailed examination of these affected individuals to undertake appropriate planning of various preventive dental and periodontal measures and facilitation of timely management of various accompanying conditions with this disease.

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Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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