Original Article

Oral health and quality of life in children with blood coagulation disorders and hemoglobinopathies. A cross-sectional study

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Abstract

Background: Blood coagulation-associated disorders including coagulopathies have been found to affect morbidity in terms of both oral and physical health. The present study aimed to assess the oral health status and quality of life in children with blood coagulation disorders and hemoglobinopathies. **Materials and Methods:** This was a cross-sectional study conducted in 'Centre for hemophilia and blood disorders'. The study group comprised of 39 children afflicted by hemophilia A, 12 children affected by hemophilia B, and 29 children having von Willebrand disease. The pediatric health-related quality of life was used for evaluating the age groups. Oral screening was performed by 2 investigators in daylight using mouth mirror and probe. Inter-examiner agreement was assessed upon 25% of studied sample using Kappa analysis and was found to be 0.89. Statistical analysis: The coded data were analyzed using the statistical software SPSS version 23.0 (IBM Corp., NY). Data entered were as frequency distribution, mean, and standard deviation. 'Chi-square' statistical tool was used for comparison of categorical variables. Continuous type of data was analyzed by 'Mann–Whitney U' test. **Results and Observations:** 67% of cases were brushing once daily whereas 33% had brushed their teeth twice a day. 78% of controls were brushing two times per day, and 22% were brushing their teeth one time daily. 78% of control subjects were brushing twice whereas 22% were brushing only once which was statistically significant ($P = 0.04$). 50% of cases had fair oral health, 30% had good, 10% were having excellent whereas 10% had poor oral health. Oral health was assessed by DMFT for permanent teeth, dmft for primary dentition, and POQL index scores. Mean ± S.D. values' comparison was found to be statistically different ($P = 0.05$ each). POQL index scores were found to have statistical significance ($P = 0.04$) in both less than 12 and 12-16 years age groups. **Conclusion:** Poor oral health was found to significantly affect the general physical health and quality of life among children affected with blood coagulation disorders and coagulopathies.

Keywords: Children, coagulation disorders, oral health, quality of life

Introduction

Blood coagulation and hemoglobinopathic disorders among children include variety of conditions ranging from benign to malignant. These diseases influence the quality of life, development, and life expectancy. These coagulation-associated

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disorders impact vascular system thus, affecting oxygen transportation, immunological defense, and blood coagulation. These coagulopathic and coagulation disorders are caused by genetic as well as environment-associated factors or both.[1] Every year, approximately three million children are being diagnosed with hemoglobinopathies.[2] Presently, 5.2 percent global population are affected with a hemoglobinopathy, out of which, 17% have thalassemia and 83% are suffering with sickle cell anemia. Most of these conditions have acquired etiology while hereditary accounts for 7% of cases.[3]

These hematological disorders may first exhibit manifestations in oral cavity, that range from mucosal alterations to severe forms of symptoms that affect quality of life specially among children. Quite often, these clinical signs may be missed resulting in delay in diagnosis as well as treatment. Advancements in medical field and hematological research have resulted in significant improvement in prognosis.

Patients suffering from hematological disorders have an increased infection risk as a response to deficient neutrophilic response and a decrease in splenic function. The administration of prophylactic antibiotic agents like amoxicillin and clindamycin has been recommended before starting any dental procedure for mitigating risk of infection.[4] Intra-venous medications like clindamycin and ampicillin are usually used as alternative ones in pre-treatment protocol before any dental surgeries in a negative medical history for vaso-occlusive event recently. However, multi-disciplinary intervention that involves communication with healthcare professionals is required in managing such patients.[5]

Hemophilic patients suffer from extended bleeding in oral cavity, especially of gingival even during eruption of tooth. A significant complication is the development of hemophilic pseudotumors in the jaw due to recurrent sub-periosteal bleeding episodes.[6] The research indicates that individuals with hemophilia have similar oral hygiene, caries prevalence, and dental care needs as those without the condition, except for increased spontaneous bleeding.[7] However, it is observed that children with hemophilia generally have poor oral hygiene and a high demand for dental care, with a reported dental caries prevalence of 73.3% and a treatment need of 93.90%.[8]

The efficient dental health care in hemophilic patients involve replacement drug therapy, anti-fibrinolytic medicinal agents, and measures for achieving local hemostasis before any surgical procedure specially, associated with inferior alveolar nerve blockage.[9] Before performing tooth extractions, both systemic as well as local therapies are required. Factor (s) replacement therapy is important for handling of moderate to severe grades of hemophilia A and B types 2 as well as type 3 von Willebrand diseases whereas systemic hemostasis inducing treatments are useful in mild cases with drug dosages adjusted to type as well as severity of disease.[10]

The surgical procedures must be planned within hospital settings in hematologists' consultations for assessment of disease severity, factor VIII inhibitor and appropriate choice of replacement therapy. In severe hemophilic individuals who do not have factor VIII inhibitors, oral procedures have been recommended post-replacement drug therapy. NSAIDs have been contraindicated in such patients due to their anti-platelet activity and increase in risk of bleeding. Herein, use of alternative agents such as paracetamol or codeine has been recommended. Intra-mucosal administration of local anesthesia has been preferred in mild-to-moderate hemophilics, in place of intra-muscular injections.[11,12]

Thus, the aim of present study was to assess the oral health and quality of life in children suffering from blood coagulation disorders and hemoglobinopathies.

Materials and Methods

Study design and setting

This cross-sectional study was conducted in a center for hemophilia and blood disorders after obtaining ethical approval from Institutional Ethical Committee (IEC23/677NM). The written consent for conducting the study was obtained from parents and assent was taken from children selected the study.

Study participants and sampling

The validity as well as reliability of studied sample was done as per the Convergent Validity of Haemo-QoL which is the instrument for assessment of Quality of life in hemophilic children and adolescents at 95% Confidence Interval and 90% power. For case study group (children with bleeding disorders and hemoglobinopathies), convenience sampling method was used. This group consisted of 39 children affected by hemophilia A, 12 children with hemophilia B, and 29 children diagnosed with von Willebrand disease. The pediatric health-related quality of life (POQL) is a tool which is used for evaluation of age groups based upon their physical as well as psychological state. POQL is 10-itemed questionnaire-based tool that has been designed for measuring oral health-related quality of life in children from both child as well as their parents' point of view.[13]

Data collection tools and technique

Data were collected under two broad headings: A) Demographic characteristics: Demographic information collected in study were 1) age, 2) gender, 3) frequency of teeth brushing, 4) general physical health, and 5) general oral health. Apart from this general physical health, oral and dental health, number of dental visits in a year, and reason for dental visit were ascertained. B) Screening of dental health: Dental health screening consisted of dmft, and DMFT indices based upon World Health Organization criteria. The screening was done by two trained clinical examiners under broad daylight using mouth mirror and probe. Inter-examiner agreement was assessed on 25% studied sample by Kappa analysis and was found as 0.89. Inclusion criteria: These were

1) Those children who were diagnosed with a bleeding disorder or hemoglobinopathy, 2) Those who consented for study participation, and 3) Those who were free from any other systemic disease. Exclusion criteria: 1) Cases where parental consent or participants' assent could not be obtained and 2) if any other systemic disease other than bleeding disorders or hemoglobinopathies was present.

Statistical analysis

The obtained data were given codes for blinded analysis. Statistical software SPSS version 23.0 (IBM Corp., NY) was employed for analyzing the data. The data were entered in form of frequency distribution, mean, and standard deviation values. Cronbach α test was used for testing validity as well as reliability of collected values. 'Chi-square' statistical test was used for comparing categorical variables. Scores of scale used and continuous measurements were tested using 'Mann–Whitney U' statistical tool.

Results and Observations

a. Demographic characteristics: On analyzing the demographic profile, it was observed that 45% of study participants were males while 55% were females whereas in the control group, there were 50% males and females participants, respectively. This was found to be statistically significant ($P = 0.05$). 67% of case group participants were brushing once a day while 33% were brushing twice daily. On the other hand, majority of control group subjects (78%) were brushing twice a day while 22% were brushing once daily. In control group, 78% were brushing twice daily while 22% were brushing once a day. This was found to be statistically significant ($P = 0.04$). On performing general physical examination, it was noted that 10% were excellent, 20% were good, 50% had fair, and 20% were having poor physical health. Whereas, 89% of control subjects demonstrated excellent, 10% had good, and 01% had fair physical health status. This was found to have good statistical difference ($P = 0.05$).

On analyzing the oral health status, 50% of case study group had fair, 30% demonstrated good, 10% had good, and 10% were in poor oral health. 89% of control subjects had excellent while remaining 11% had good oral health. This was found to have good statistical significance ($P = 0.05$).

On studying the number of dental visits, it was seen that 30% of affected subjects visited dental six months back, 41% had a dental visit one month back, 09% had visited dentist 3 months back, however, a large numbers of patients (40%) had never visited a dental set-up. On the other hand, 52% of the control group subjects were found to have undergone dental procedure six months back, 24% visited dentist one year back, 165 had a dental visit three months previously, and 8% had never visited a dental clinic. This was found to be statistically significant ($P = 0.03$) [Table 1 and Graph 1].

b. Oral health status and POQL index: The oral health status was determined by analyzing the DMFT (permanent teeth), dmft (primary dentition), and POQL index scores. On comparing the mean ± S.D. values between case and control subjects, statistically significant differences ($P = 0.05$ each) were found. On studying the POQL index scores, statistically significant difference ($P = 0.04$) were obtained in both <12 and 12 to 16 years age groups [Table 2 and Graph 2].

Discussion

The hereditary forms of coagulopathies constitute diseases that result from quantitative or qualitative deficiencies of platelets and/or coagulation factors. These diseases can be characterized by hemorrhage of variable severity that may have spontaneous origin or may be associated with traumatic episodes. These patients require transfusion of blood derived components or their derivatives. Transfusion of replacement factor therapy is done in conditions that involve bleeding episodes or as prophylactic drug therapy.[13]

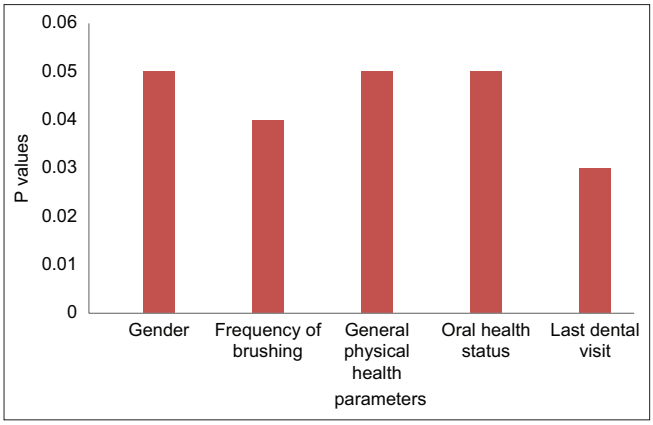
These patients face difficulty in availing treatment at primary healthcare centers. There are dedicated healthcare centers that include dental practitioners in the medical team thus, making dental care accessible to these individuals.

Table 1: Table demonstrating sociodemographic features of studied group and control subjects

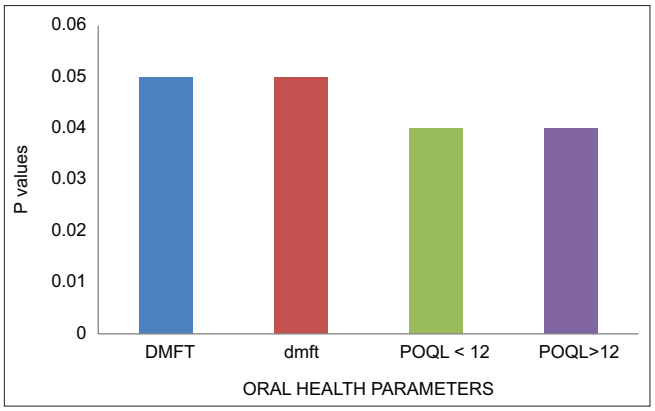
Parameters	Case group	Control group	*P*
Gender			
Male	45%	50%	0.05
Female	55%	50%	
Frequency of brushing			
Once a day	67%	22%	0.04
Twice a day	33%	78%	
General physical health			
Excellent	10%	89%	0.05
Good	20%	10%	
Fair	50%	01%	
Poor	20%	0%	
Oral health status			
Excellent	10%	89%	0.05
Good	30%	11%	
Fair	50%	0%	
Poor	10%	0%	
Last dental visit			
3 months back	09%	16%	0.03
6 months back	30%	52%	
One year back	41%	24%	
Never	20%	08%	

Table 2: Table showing dmft, DMFT and POQL index scores in case study and control groups

Indices	Mean±S.D		*P*
	Control group	Case group	
DMFT index	2.3±0.21	7.1±2.3	0.05
dmft index	1.9±0.13	5.6±0.23	0.05
POQL index			
<12 years	16.1±0.16	35.2±21.6	0.04
12-16 years	12.6±0.45	38.2±21.4	0.04



Graph 1: Demonstrating *P* values of studied socio-demographic parameters



Graph 2: Demonstrating *P* values of studied oral health parameters

The oral health in a child exhibits a direct impact over mastication, speech, expression, and physical appearance. Pain originating from oral problems negatively influences an affected child's day-to-day life.[14] Oral health-related quality of life is a multi-dimensional tool mainly used for assessment of one's perceptions about a disease and/or condition.[15]

Amirabadi *et al.*[15] (2019) in their observations reported significant inter-relationship between dental caries and oral health-related quality of life in thalassemia major afflicted children. Additionally, a decline in OHRQoL was found to be related with an increase in issues affecting oral health. Ebeid *et al.*[16] (2020) LSO reported a negative influence of thalassemia over OHRQoL.

The present study assessed the oral health status and quality of life of children afflicted with bleeding and coagulopathic disorders. It was observed that there were higher percentages of affected females with this group of disorders. The frequency of brushing was found to have statistically significant difference with control group subjects. Similarly, other sociodemographic parameters also were found to have statistically significant differences [Table 1 and Graph 1]. Oral health status was evaluated by means of indices like dmft, DMFT, and POQL which were also found to have statistically significant [Table 2 and Graph 2].

De Silva *et al.*[17] (2024) in their study reported poor correlation of OHRQoL with oral hygiene (ρ = - 0.383 and P = 0.004) and gingival health (ρ = -0.327 and P = 0.014). Higher untreated cases of dental caries were found to be associated with poor oral health-related quality of life (P = 0.009). Poor status of oral health in children as well as adolescents has been found to have significant negative impact over one's overall quality of life.

Yazıcıoglu *et al.*[18] (2022) in their results derived from POQL demonstrated that hemophiliac patients have poor oral health-related quality-of-life (OHR-QoL) when compared to control subjects. Higher total as well as sub-groups POQL scores were obtained in hemophilia patients that was suggestive of poor OHR-QoL and DMFT scores. Similarly, oral health-related treatment needs and urgency for treatment were found to be higher in hemophiliacs than healthy controls.

In a similar study conducted by Baskirt *et al.*[19] (2009) patients suffering from hemophilia were found to have requirement for dental treatment.

Hartl *et al.*[20] (2013) in their study conducted on severe patients of hemophilia reported worst quality of life scores in terms of physical functions general physical and oral health.

Fadel *et al.*[21] (2020) in their study reported high dental caries experience in their study on thalassemic patients, however, they found no negative effect over their quality of life which is in contrast to our study findings.

Treviño-Tijerina *et al.*[22] (2023) conducted an online survey in patients with hematological diseases and found that disorders like leukemia, anemia, hemophilia, and thalassemia not only pose significant challenges to children's general health but also have a substantial impact on their oral health.

Fernandes *et al.*[23] (2016) investigated the influence of sickle cell disease, socioeconomic characteristics, and oral conditions on oral health-related quality of life of children and teens. A significant difference was observed in the oral health-related quality of life between adolescents with sickle cell disease and controls only in relation to malocclusion.

Trindade GC *et al.*[24] (2019) evaluated the Qol of patients with hemophilia using the WHOQOL-bref and the Haemo-A-Qol instruments and concluded that hemophilia had a higher negative impact upon physical aspects, sports, and leisure in the patients of the sample.

Limitations

The major limitation of the present study was the small size of population studied due to which complete representation of each type of disease such as hemophilia A, B, thalassemia, etc.,

could not be assessed. Hence, studies with longer duration or more sample size distribution must be done.

Conclusion

Bleeding disorders and coagulopathies have been found to influence oral health status of affected children. Due to presence of poor oral health, it affects the overall quality of life and general health. The present study highlighted the requirement for raising awareness among parents, care-givers as well as dental professionals regarding treatment of such children.

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Conflicts of interest

There are no conflicts of interest.

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