10.5005/jp-journals-10063-0001

ORIGINAL ARTICLE

Occurrence of Dental Abnormalities in Hemophilic Patients in the City of Davangere, Karnataka, India

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ABSTRACT

Aim: This study was carried out to determine the level of awareness about the importance of oral health in hemophilic children and their caretakers as well as to examine the oral condition in children with hemophilia and compare them to general population.

Materials and methods: A descriptive cross-sectional observational study was conducted in the city of Davangere. The study consists of total 100 children which were divided into group I —children having hemophilia (n = 50) and group II—normal healthy children (n = 50) of age 3 to 18 years. The oral cavity of both the groups was examined to assess to detect presence of any hard and soft tissue anomalies. By interviewing the parents of hemophilic patients, their demographic data, family history, and knowledge and understanding of value of oral health were assessed. Data were analyzed by chi-square and student's unpaired t-test.

Results: No significant difference in the presence of oral anomalies in both the groups was observed.

Conclusion: The study concluded that there is no major difference in the prevalence of dental abnormalities in hemophilic and healthy children. Also, educating the mass about oral health and maintenance is as important as treating the hemophilic patient for oral diseases.

Keywords: Dental abnormalities, Hemophilia, Oral hygiene.

How to cite this article: Nagaveni NB, Yadav S, Poornima P, Bharath KP, Mathew MG, Shruthi AS. Occurrence of Dental Abnormalities in Hemophilic Patients in the City of Davangere, Karnataka, India. CODS J Dent 2016;8(1):1-5.

Source of support: Nil

Conflict of interest: None

INTRODUCTION

Hemophilia also known as the "Royal Disease" is one of the most commonly occurring hemorrhagic diathesis across the globe with an occurrence of 1 per 10,000 people. Among all the congenital bleeding disorders, it accounts for 80% of bleeding disorders.[1] Hemophilia is a long-known disorder. Hemophilia is been long-known since 2nd century mentioned by Jewish Rabbinical writing and an Arabian physician from Cordoba named Albucasis of 12th century AD, wrote of males in particular village who had died of uncontrollable bleeding. Later, it gained popularity as "Royal Disease" as it ran through the generation of royal families of England, Spain, Germany, and Russia. Queen Victoria of England was a carrier of the hemophilia gene and subsequently passed on to several royal families. Dr John Conrad Otto in 1803 was the one to first recognize the hereditary pattern and male predominance of the disorder.[2] Due to its deliberating condition, a patient suffering from this disorder should be considered as a special case.

The oral tissue is blessed with enormous blood supply and become a hot spot for hemorrhage in this group of patients.[2] Hard tissue anomalies, such as enamel hypoplasia or severe fluorosis increases the chances of enamel been eroding off and exposing the vascular pulp tissue. Various hard or soft tissue pathologies and treatment of oral cavity may predispose such individual to risk of bleeding excessively. The dentists are at a good position to identify the condition first and take a timely measure to prevent further deterioration which might demand for more invasive procedure.[2] A prior knowledge about difference in the oral health seen in hemophilic patient would make the dentist more vigilant access the condition and quick to take the necessary steps.

Thus, the purpose of this study is to examine different aspects of dental health or disease in children with hemophilia and to compare them to matched controls from the general population. Also, the study aimed to determine the level of awareness of value of oral health in caregivers of hemophilic patient.

MATERIALS AND METHODS

A descriptive cross-sectional study was conducted by the Department of Pedodontics and Preventive Dentistry, College of Dental Sciences, Davangere, Karnataka, India. An ethical clearance was obtained from the Ethical Committee of College of Dental Sciences, Davangere.

Two-stage sampling procedure was adopted to select sample. In the first stage, 50 children with hemophilia

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and normal children were selected randomly from the Hemophilia Society, Davangere, Karnataka and from the Department of Pedodontics, College of Dental Sciences, Davangere, Karnataka, India respectively. Probability proportional to the size was used to ensure that all would have an equal chance to be selected in the study. In the second stage of sampling, children aged 3 to 18 years were selected from each group.

The 50 hemophilic children suffering from hemophilia were enrolled names, residential address, parental occupation, and other criteria in Karnataka Hemophilia Society for general treatment and follow-up. These children were analyzed and compared to 50 healthy children who visited to the Department of Pedodontics in the same age and gender distribution. These children were resident of in and around Davangere city. Consent was obtained from the parent of the children at the commencement of investigation.

The patients were seated on an ordinary chair under natural illumination in a room using a mouth mirror and probe. To improve the visibility, prior to the examination, each child was asked to rinse their mouth with water, the teeth were then dried using cotton rolls. Both primary and permanent dentition was taken to consideration in all children. A survey pro forma was created to gather data from the sample. All the examinations were carried out by a single qualified examiner and parameters examined were:

- Hard tissue anomalies—presence of supernumerary teeth, enamel hypoplasia, fluorosis, and traumatic injuries.
- Soft tissue anomalies—presence of ulcers or any other changes on tongue, buccal mucosa, floor of the mouth, and lips.

Inclusion Criteria

- Study subjects should be aged between 3 and 18 years.
- Only boys were included.

Exclusion Criteria

- Children with deviated nasal septum, enlarged adenoid, and clefts of the lip/palate.
- Children with other systemic disorders, such as respiratory disorders, neuromuscular, cardiac disorder, etc.

The response obtained was tabulated and the results were expressed as frequency distributions and computed in percentages. The observations were statistically analyzed using the chi-square test, student's unpaired t-test.

RESULTS

Out of 50 children, factor VIII deficiency was seen in 35 children, whereas 15 children shown deficiency of factor IX (Table 1). A total of 18 children had the positive family history, out of which 14 (28%) children had their mother's brother been affected by hemophilia, whereas 4 (8%) children had their own brothers affected. About 32 (64%) children had a negative family history with none of the family member suffering by hemophilia (Table 2). When the subject were compared according to the age in both the groups, 10 and 9 subjects of hemophilia and control respectively fell in the age group of 3 to 6 years, 29 and 31 subjects of hemophilia group respectively fell in the age group of 12 to 18 years. The mean age score was 9.72 ± 3.85 and for control group it was 9.58 ± 3.10 with a t-value of 0.02 and p-value 0.084, thus, there was no statistical difference in age (Table 3).

Out of 50 children examined in study group, parents of 36 children had knowledge and awareness regarding oral hygiene and parents of 14 children did not have the knowledge and awareness regarding oral hygiene. Whereas in control group, parents of 22 children had knowledge and awareness regarding oral hygiene and parents of 28 children did not have the knowledge and awareness regarding oral hygiene (Table 4). When the anomalies, such as supernumerary teeth, enamel hypoplasia, fluorosis, and trauma were considered again, there was no statistical significant difference found between the groups. Supernumerary tooth was seen in 2% of the subjects in the test group only. Enamel hypoplasia was

Table 1: Distributions of study group according to the type of hemophilia

Factor	*Number of patients affected*	*Percentage*
VIII	35	70
IX	15	30
PTH	0	0

PTH: Parathyroid hormone

Table 2: Affected family member

Affected family member	*Number of patient*	*Percentage*
Mother's brother	14	28
Brother	4	8
None	32	64

Table 3: Agewise comparison of test and control group

Age (years)	*Hemophilia group (n = 50)*	*Control group (n = 50)*
3–6	10 (20%)	9 (18%)
6–12	29 (58%)	31 (62%)
12–18	11 (22%)	10 (20%)
Mean ± SD	9.72 ± 3.85	9.58 ± 3.10

Table 4: Knowledge and awareness of parents regarding maintenance of oral hygiene in both the groups

Awareness	*Hemophilia group (n = 50)*	*Control group (n = 50)*
Yes	36 (72%)	22 (44%)
No	14 (28%)	28 (56%)

Table 5: Comparison of dental anomalies among test and control group

Parameters	*Hemophilia group (n = 50) Mean ± SD*	*Control group (n = 50) Mean ± SD*	*Significance*	*Chi-square test*
Supernumerary teeth				
Present	1 (2%)	0 (0%)	0.315	χ^2 – 1.010
Absent	49 (98%)	50 (100%)		df – 1
Enamel hypoplasia				
Present	5 (10%)	3 (6%)	0.461	χ^2 – 0.543
Absent	45 (90%)	47 (94%)		df – 1
Fluorosis				
Present	1 (2%)	2 (4%)	0.558	χ^2 – 0.334
Absent	49 (98%)	48 (96%)		df – 1
Trauma				
Present	5 (10%)	4 (8%)	0.727	χ^2 – 0.122
Absent	45 (90%)	46 (92%)		df – 1

df: Degree of freedom; SD: Standard deviation

Table 6: Comparison of soft tissue among test and control group

Parameters	*Hemophilia group (n = 50) Mean ± SD*	*Control group (n = 50) Mean ± SD*	*Significance*	*Chi-square test*
Tongue (ulcer)				
Present	1 (2%)	1 (2%)	1.000	χ^2 – 0.000
Absent	49 (98%)	49 (98%)		df – 1
Buccal mucosa (ulcer)				
Present	1 (2%)	0 (0%)	0.315	χ^2 – 1.101
Absent	49 (98%)	50 (100%)		df – 1
Floor of mouth (ulcer)				
Present	0 (0%)	0 (0%)	–	–
Absent	50 (100%)	50 (100%)		
Lips (ulcer)				
Present	3 (6%)	2 (4%)	0.646	χ^2 – 0.211
Absent	47 (94%)	48 (96%)		df – 1
Other changes				
Present	7 (14%)	7 (14%)	1.000	χ^2 – 0.000
Absent	43 (86%)	43 (86%)		df – 1

df: Degree of freedom; SD: Standard deviation

seen in 10 and 6% of the subjects of test and control group respectively. Fluorosis was seen in 2 and 4% of the subjects of test and control group respectively. The incidence of trauma was seen in 10 and 8% of the subjects of test and control group respectively (Table 5). There was an ulcer present on the dorsum surface of tongue in 2% of the subjects in both the groups. Buccal mucosa also showed an ulcer in 2% of the subjects in test group only. When floor of the mouth was considered, there were no such findings in both the groups. There was a lip ulcer which was seen in 6 and 4% of the subjects of test and control group respectively. Other soft tissue changes, such as gingivitis was seen in 14% of the subjects of both the groups (Table 6).

DISCUSSION

Hemophilia in actual sense is a group of hereditary genetic disorder that impairs the body's ability to control bleeding, in case there is a breach in the blood vessel. Hemophilia can be classified as hemophilia A, B, and C based on the deficient clotting factor. Deficiency of factor VIII is seen in hemophilia A (antihemophilic globulin),[2] whereas in hemophilia B and C, factor IX and Plasma thromboplastin antecedent is deficient respectively.[3]

The etiology for this condition is a sex-linked recessive transition of affected X chromosome which causes a deficiency of factor VIII that can be either complete or partial. Hemophilia A arises from a variety of mutations; some 150 point mutations have been characterized. It is transmitted by asymptomatic female carriers and mostly males are the sufferers because the genetic constitution of male shows presence of single X chromosome, which, if affected, results into clinical manifestations. On the contrary, if such condition prevails in a female, is masked by the other normal X chromosome and hence female becomes carriers and not sufferers. The condition manifests in alternative generation where the defective gene is passed from grandfather to daughter and then to her son. Based on plasma level of factor VIII, hemophilia

was classified as severe (<1%), moderate (1–5%), and mild (>5%).[4] The positive inheritance was noted in this study. The questioners were asked regarding the same, out of 50 patients, 18 (36%) patients had a positive family history where their own brothers or mother's brother were affected. While in 32 (64%) patients gave a negative history with none of the other family member affected. This was not in accordance to the established facts and the reason could be unawareness of the family or an incorrect answer to the questions.

Noting the epidemiology of hemophilia, USA ranks first in the prevalence of hemophilic cases[5] which is 13,276 and 1 per 5,032 male births according to Soucie et al.[6] Followed by India (11,586), Brazil (8,839), China (8,921), and the UK (5,424). According to the study conducted by Kar et al,[7] India has the third largest number of patients with bleeding disorders and second highest number of global patients with hemophilia A. The count of reported cases is 11,586 while the estimated prevalence could be around 50,000 patients.

Among all types of hemophilia, the prevalence of hemophilia A is maximum, i.e., 1 per 10,000[2] followed by hemophilia B which affects only 1 per 300,000 males born alive.[1] In this study, out of 50 hemophilic children 35 (70%) had hemophilia A and the rest, i.e., 15 children had hemophilia B (15%). Hemophilia C which is a rare phenomenon was not seen in the study group. Just like this study, the epidemiological survey of Kar et al also showed that out of all hemophilia cases 70% suffered from type A.[7] Study of Ahmad et al[8] and Chuansumrit et al[9] showed hemophilia A was the most common and was seen in 52.31% and 91.3% respectively. Similar results were seen in the review of Zaliuniene et al[10] which showed hemophilia A occurs in 80 to 85% of patients and has an incidence of 1:5,000 in the male population, whereas hemophilia B (or Christmas disease) diagnosed 10 times less frequent than hemophilia A. It accounts 10 to 15% of hemophilia cases with the incidence of 1:15,000.

A review of the literature regarding the care of hemophilic patients showed a paucity of material on this important subject and lack of information on the dental health of this group of patients. [11] Oral pathologies and its treatment, both stand as a risk to excessive bleeding in hemophilic patients.[3] Till date, the focus of dentistry has been dental extractions rather than conservation of the dentition and promotion of oral health.[1] Preventative dentistry may help to forbid the requirement of extensive oral rehabilitation and avoid complications. However, for implementation this idea as an awareness of the importance of oral health among hemophilic patient and parents and understanding of the predisposing features that can led to dental problem by dentist is vital. Therefore, this study also evaluated the knowledge and awareness among the parents of hemophilic children about the importance of oral hygiene. When questions were asked to parents regarding the knowledge and awareness of oral hygiene and oral bleeding, parents of 36 (72%) patients gave a positive response. In this study, 72% of parents answered positively about the awareness of dental health and their main concern was with bleeding in during the time of eruption and shedding of the teeth. In the city of Republic of Iran, the study conducted by Abrisham et al[12] was in accordance with this study. The questionnaire answered by the caretakers of hemophilic patients of Iran reflected their fair knowledge of importance of oral hygiene. This was in contrast to the finding of Kabil et al[13] whose survey revealed that the children and their caregivers know very little about oral health and an oral needs, assessment has never been conducted on them. As a dentist, it becomes our duty to educate and give instruction to the general population about the measures to improve the oral condition and its effect on the general health. Such efforts might make the patient realize the need to return regularly for professional prophylaxis, examination, and treatment and adoption of the preventive steps, such as the use of fluoride. These facts hold true for all individuals; however, the issue gains more importance as dental care affects the general health of hemophilic patients. Also, confronted with specific complexities of the disease, the families with hemophilic patients do not pay sufficient attention to their dental care. Another reason for the ignorance can be the fear of bleeding during the procedure. Either of them results in conditions, such as dental caries and periodontal disease which would then require more invasive treatment. Thus, proper knowledge of the subject is also necessary for these patients. Pashapoor and Gol Mohammad Loo focused on the positive role of instruction in hemophilic patients.[14]

According to Zaliuniene et al,[10] 14% of all hemophilia patients have been first diagnosed following an episode of severe oral bleeding, following tooth brushing, food abrasion, or with periodontal disease due to the number of enlarged capillaries close to the surface of the thinner regions of the gingiva. The most common site of oral bleeding was the labial frenulum and the tongue. This puts the dentist in the first place to chance diagnose the condition.[10] However, this study included a set of young children and history of oral bleeding mainly during the time of eruption/shedding of primary teeth, followed by trauma-induced bleeding especially in the lips and tongue region. Similar observation was made by Salem,[15] who mentioned that 55% patients gave the history of oral bleeding mainly during the time of eruption/shedding of primary teeth, followed by trauma-induced bleeding especially in the tongue region.

Considering the presence of oral hard tissue anomalies in the hemophilic patient, this study included the oral examination to note the presence of any supernumerary teeth, enamel hypoplasia, fluorosis, and trauma. However, when compared to control group, none of these showed a statistically significant difference. This was also supported by Sonbol et al[16] study in which they said that the white enamel opacities were detected only in permanent dentition and there was no significant difference in the number observed between hemophilia and control groups. There were no developmental enamel defects in the primary teeth in this investigation. Similar findings were mentioned in the study of Salem[15] where hypoplasia of first permanent molar as was observed with no difference between groups and no other anomalies of shape, size, or color of teeth were found according to clinical examination. Also, the survey of Rajantie et al[17] showed only 43% of patients having developmental enamel defects.

The study conducted had a drawback of limited sample size and therefore the results cannot be generalized. Further studies with larger group of subjects and new approaches to assess the children with hemophilia compared with healthy controls are needed.

CONCLUSION

In a country like India, hemophilia is not given the priority it deserves as these areas have high numbers of other serious health problems. However, preventive measure, such as educating general public, understanding among dentist about the condition of oral condition in hemophilic patients is the key importance to the population's health and is necessary for targeting patients with hemophilia for primary oral health prevention as well as for implementing timely secondary prevention, both reducing dental treatment needs and their related risks in this vulnerable segment of population.

REFERENCES

1. Mythri H, Jagdish GR, Chandu GN. Dental caries experience among haemophilic children. RJPBCS 2013 Oct-Dec;4(4): 1233-1238.
2. Kumar JN, Kumar RA, Varadarajan R, Sharma N. Specialty dentistry for the hemophiliac: is there a protocol in place? Indian J Dent Res 2007 Apr-Jun;18(2):48-54.
3. Saxena S, Shashikiran ND. Prevalence of dental caries and treatment needs among hemophilic children of Kota city, Rajasthan. Annals and Essences of Dentistry 2010 Apr-Jun;2(2):18-21.
4. Madan N, Rathnam A, Bajaj N. Treatment of an intraoral bleeding in hemophilic patient with a thermoplastic palatal stent — A novel approach. Int J Crit Illn Inj Sci 2011 Jan;1(1): 79-83.
5. World Federation of Hemophilia (WFH). 29. Report on the Annual Global Survey 2011. Canada: WFH; 2013.
6. Soucie JM, Evatt B, Jackson D. Occurrence of hemophilia in the United States. The hemophilia surveillance system project investigators. Am J Hematol 1998 Dec;59(4): 288-294.
7. Kar A, Phadnis S, Dharmarajan S, Nakade J. Epidemiology & social costs of haemophilia India. Indian J Med Res 2014 Jul;6140(1):19-31.
8. Ahmad F, Kannan M, Ranjan R, Bajaj J, Choudhary VP, Saxena R. Inherited platelet function disorders versus other inherited bleeding disorders: an Indian overview. Thromb Res 2008;121(6):835-841.
9. Chuansumrit A, Mahasandana C, Chinthammitr Y, Pongtanakul B, Laossombat V, Nawarawong W, Lektakul Y, Wangruangsatid S, SriboriboonsinL, Rojnakarin P, et al. National survey of patients with hemophilia and other congenital bleeding disorders in Thailand. Southeast Asian J Trop Med Public Health 2004 Jun;35(2):445-499.
10. Zaliuniene R, Aleksejuniene J, Peciuliene V, Brukiene V. Dental health and disease in patients with haemophilia—a case-control study. Haemophilia 2014 May;20(3):e194-198.
11. Nagaveni NB, Arekal S, Poornima P, Hanagawady S, Yadav S. Dental health in children with congenital bleeding disorders in and around Davangere: A case-control study. J Indian Soc Pedod Prev Dent 2016 Jan-Mar;34(1):76-81.
12. Abrisham M, Tabrizizadeh M, Ghateh A. Knowledge of orall hygiene among hemophilic patients referred to Iranian Hemophilia Society. J Dent Res Dent Clin Dent Prospects 2009 Spring;3(2):60-63.
13. Kabil N, ElAlfy MS, Metwalli N. Evaluation of the oral health situation of a group of Egyptian haemophilic children and their re-evaluation following an oral hygiene and diet education programme. Haemophilia 2007 May;13(3):287–292.
14. Pashapoor N, Gol Mohammad Loo S. The acceptance of self-treatment in hemophilic patients after receiving instructions. J Med Res Sci. 1998;5:140-142.
15. Salem K, Eshghi P. Dental health and oral health-related quality of life in children with congenital bleeding disorders. Haemophilia. 2013 Jan;19(1):65-70.
16. Sonbol H, Pelargidou M, Lucas VS, Gelbier MJ, Mason C, Roberts GJ. Dental health indices and caries-related microflora in children with severe haemophilia. Haemophilia. 2001 Sep;7(5):468-474.
17. Rajantie H, Alapulli H, Mäkipernaa A, Ranta S. Oral health care in children with haemophilia in Helsinki, Finland. Eur Arch Paediatr Dent 2013 Oct;14(5):339-343.