



## Dental Management of 6 Year Old Child with “Moderate Hemophilia A.”: A Case Report

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Date of Submission: 20-07-2023

Date of Acceptance: 31-07-2023

### ABSTRACT

Hemophilia is one of the hereditary bleeding disorders, caused by deficiency of one or more clotting factors. It is classified as hemophilia A, B and C which occur due to deficiency of factor VIII, IX and X respectively. Hemophilia A is X linked recessive hereditary disorder which is most common of the three. Systemic conditions like hemophilia raises a concerns about dental treatment which routinely consist the use of sharp dental hand instruments & rotary instruments which addresses highly vascular intraoral soft tissue. This case report describes chair side dental treatment of 6 years old child, who was a known case of moderate hemophilia-A.

**KEYWORDS:** Hemophilia A, Pediatric Patient.

### I. INTRODUCTION

Hemostasis is a defense mechanism which protects the body from losing blood after a vascular injury.<sup>(1)</sup> Hemophilia A and B are the main inherited bleeding disorders linked to the X chromosome which arise due to mutations in the genes for factor VIII and IX causing deficiency or a functional decline of these proteins in plasma.<sup>(2)</sup> Their frequencies are 1 in 5,000 and 1 in 30,000 male live births, respectively.<sup>(3)</sup> (Figure 1).

The symptoms depend on the deficiency degree in the clotting factor and are classified as severe, factor levels <1%; moderate, factor levels of 1-5%, and mild, factor levels >5% (table 1).<sup>(4)</sup> Clinical manifestations of severe forms are characterized by spontaneous bleeding. Moderate forms seldom have spontaneous bleeding and only mild forms are associated with trauma or invasive procedures without having received replacement treatment.

Common sites of bleeding include joints, muscles, and skin. Hemarthroses (joint hemorrhages) are frequently observed, with symptoms including pain, stiffness, and limited

range of motion.<sup>(4)</sup> Repeated episodes of hemarthroses or muscle hemorrhage result in chronic musculoskeletal disease and culminate in debilitating painful arthritis and disability.<sup>(4)</sup> Pseudo tumors (hemorrhagic pseudocysts) may occur in several locations including the jaw, in which case curettage is indicated. With all severities of hemophilia A common cause of bleeding in children are mouth lacerations, during the time of eruption or shedding of primary teeth and even without discernible trauma.<sup>(4)</sup>

The definitive diagnosis of suspected hemophilia must be done through clinical and family history along with laboratory tests like a flow cytometry, bleeding time (BT), activated partial thromboplastin time (APTT), prothrombin time (PT) and thrombin time (TT).<sup>(5)</sup> Treatment involves replacing the missing clotting factor. The amount of factor VIII concentrate needed depends on bleeding severity, the site and patient's body built.<sup>(2)</sup>

This case report demonstrates the safe and successful chair side delivery of full dental rehabilitation with multiple restorations through efficient teamwork management and treatment options that minimized the risk of perioperative bleeding.

### CASE

A six year old male patient, a known case of Hemophilia A, who was under regular care of pediatrician in NKPSIMS medical college was referred to the department of Pedodontics with the complaint of decayed teeth in upper and lower jaw. Upon enquiring his mother for medical history, family history for hemophilia was present, maternal uncle was suffering from hemophilia. Patient was diagnosed with moderate hemophilia- A at the age of 3 years. Previous history of periodic joint pain, surgical excision of scalp hematoma 1 month back was carried out. Patient was on factor VIII therapy



periodically every 2 months. Previous dental history of exfoliation of 81 which was associated with severe bleeding for which patient was hospitalized and factor replacement therapy was done before 15 days.

Intraoral examination showed presence of multiple carious primary teeth. Deep Occlusal caries was present with 75 and 85. Multisurface caries was prominent with 84 and 74. Arrested caries was seen with 64. Mesial caries was seen with 51, 61 & 52 and occlusal pit caries with 55 was seen. Intraoral periapical radiographs were taken which confirmed the multiple carious lesions. Radiolucency involving enamel, dentine and approaching towards pulp seen with 74,75 and 84, 85 suggestive of moderate caries. Based on clinical and radiographic findings final diagnosis: reversible pulpitis with 74,75 and 84,85 was confirmed.

Pre-operative blood investigation values were as follows: Hemoglobin - 12.5 gm%, prothrombin time -13 seconds, and activated partial thromboplastin time (aPTT) - 48.22 sec (Normal – 26.7sec), FACTOR VIII-2.5% OF NPP (Normal : 50-150% OF NPP).

After medical fitness examination by pediatrician, a treatment plan was prepared. Treatment plan was explained to parents and informed consent was obtained.

### TREATMENT.

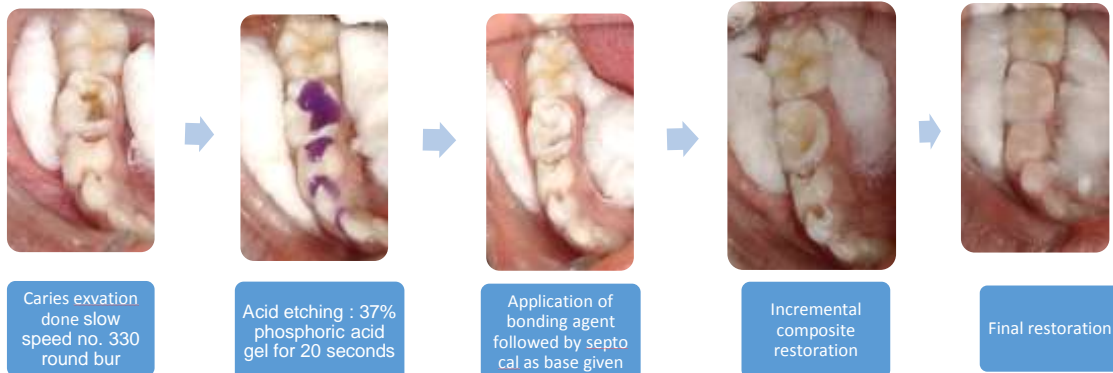
Oral antifibrinolytic agent Tranexamic acid 250 mg TDS (Tranexa<sup>®</sup>) was started before 24 hrs of treatment. Before 2 hours of treatment

Factor VIII was administered intravenously. Treatment was done in as minimum appointments as possible, caries excavation was done by spoon excavator initially and cavity modification done by high speed airotherhandpiece, extra care was taken to protect the tongue, buccal mucosa and lips. Conservative extensions of gingival margins were prepared, as it will ease the concern of soft tissue injury. Wet cotton rolls were placed to reduce mucosal bleeding during cavity preparation. During Composite restoration isolation was achieved by cotton rolls and saliva ejectors. The coronal enamel was then etched for 20 seconds, rinsed with water and air dried followed by application of bonding agent which was then lightcured, A2 shade of composite was selected followed by restoration with incremental method (Nano hybrid composite material (Hercules precise) and light cured for 60seconds.

Treatment was completed in two appointments or stages as follows: on the first appointment, caries excavation was done. Treatment was done in two appointments or stages as follows: on the first appointment, caries excavation was done with maxillary anteriors (51,61,62) along with maxillary (64) & mandibular molars (74,75) followed by composite restoration. In 2<sup>nd</sup> appointment, Caries excavation was done with maxillary (55) & mandibular molars (84,85) followed by composite restoration.

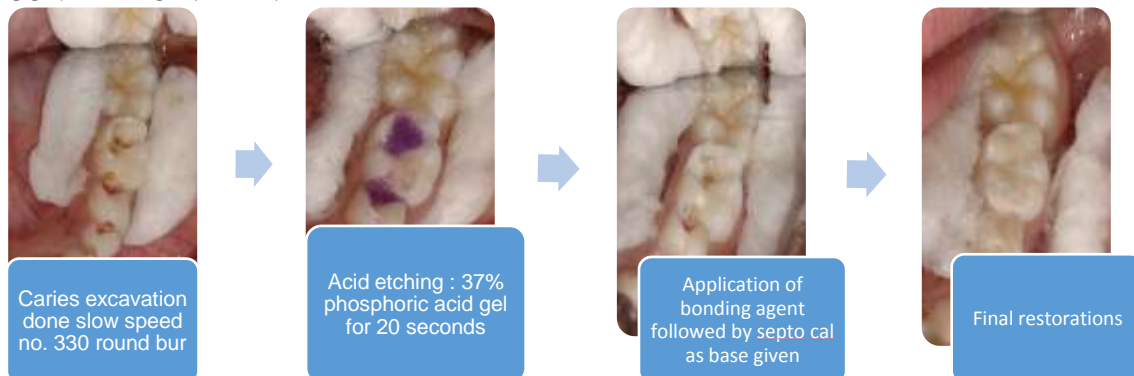
High speed vacuum suction was used with care. If bleeding occurred due to laceration of soft tissues topical application of Tranexamic acid was done.

### FIRST APPOINTMENT





## SECOND APPOINTMENT



## II. DISCUSSION

In hemophilic children, the highly vascular oral cavity is frequent for hemorrhage episodes. Patients are prone to bleed spontaneously in cases of mouth lacerations, during the time of eruption or shedding of primary teeth and even without discernible trauma.<sup>(1)</sup> Nagaveni et al (2016) reported that higher DMFT & DMFS index resulted in children with hemophilia was a consequence of neglected or insufficient tooth brushing. When caries are considered, children with hemophilia must be seen as a high-risk group.<sup>(6)</sup>

In present case whole treatment is completed in 2 appointments. As per Guidelines for the Management of Hemophilia given by World Federation of hemophilia recommended that ; most patients with bleeding disorders are to receive routine regular outpatient dental care.<sup>(3)</sup> Appointments should be arranged so that maximum treatment is accomplished per visit in order to minimize the need for unscheduled factor infusions. For patients with moderate and severe haemophilia A and B, coagulation factor replacement therapy is the main form of therapy.<sup>(3)</sup> The timing of administration is important as factor levels will decline, therefore dental procedures should be performed as close to the time of administration of factor concentrate as possible, normally within 30 minutes to an hour. Factor replacement therapy may be prescribed on a prophylactic basis to prevent bleeds, or may be administered 'on-demand' when a bleed occurs. It is important to take into account these guidelines for management and dental treatment of these patients.<sup>(7)</sup> In invasive procedures for hemophilia A, it is recommended to raise the deficient factor to 100%. In less invasive procedures, it is required to raise the factor of 40 to 60% one hour before starting the procedure.

Most restorative procedures on primary teeth are successfully completed, without factor concentrate replacement.<sup>(4)</sup> Saliva ejectors must be

used with caution to prevent sublingual hematomas.<sup>(4)</sup> Care must also be used in the placement of intraoral radiographic films, particularly in highly vascular sublingual tissues.

Tranexamic acid (Cyklokapron®) competitively inhibits the activation of plasminogen to plasmin thereby inhibiting fibrin clot lysis. Orally, tranexamic acid given at a dose of 15–25 mg/kg which approximates to 1 g for every 6-8 hours. Ideally this should be given two hours pre-operatively.<sup>(8)</sup> Tranexamic acid is freely soluble in water and it has been suggested that a 500 mg tablet could be crushed and dissolved in water and apply topically.<sup>(9)</sup> Proper toothbrushing, flossing, topical fluoride application & adequate systemic fluoride administration at regular intervals is an effective approach to the prevention of dental problems.<sup>(9)</sup>

## III. CONCLUSION

Hemophilic patients form a priority group for dental and oral health care, since bleeding after dental treatment may cause severe or even fatal complications. Moreover, maintenance of oral hygiene and prevention of dental diseases is of great significance to improve the quality of life and avoid the dangers of surgery.

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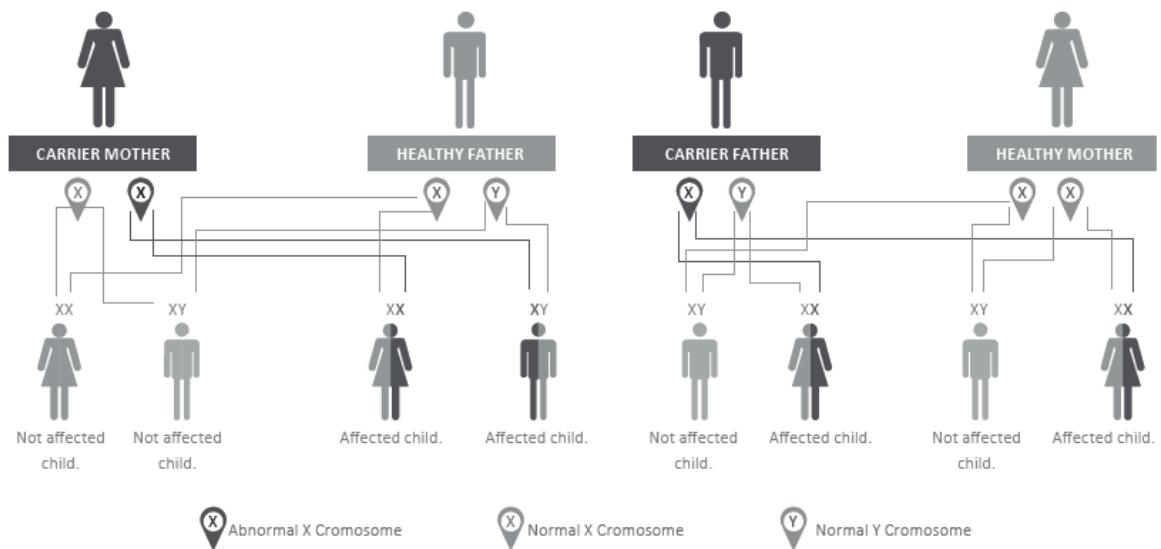


Figure 1. Inherited hemophilia patterns. Source: World Federation of Hemophilia 2008

DISEASE SEVERITY.	CLOTTING FACTOR LEVEL (VIII or IX).	HEMORRHAGIC EPISODES.
Severe	<1% (0.01 UI/ml)	Spontaneous and especially in muscles and joints.
Moderate	1-5% (0.01-0.05 UI/ml)	Occasionally spontaneous. Severe with trauma or surgery.
Mild	5-40% (0.05-0.040 UI/ml)	Only with trauma or surgery.

Source: World Federation of Hemophilia 2008.

Table 1. Classification of disease severity



Fig 2 : Front View



Fig 3 & 4 pre-operative



Fig 5-6 : Post-operative