

## Case Report

## Dental Management for a Child with Hemophilia A: A Case Report

Mohammed Saeed Aldossary\*

Postgraduate student, Department of Pediatric Dentistry, The University of Edinburgh, Edinburgh Dental Institute, Lauriston Place, Edinburgh EH3 9HA, United Kingdom (Currently: Specialist in Pediatric Dentistry. Ministry of Health, Riyadh, Saudi Arabia)

**\*Corresponding Author:**

Mohammed Saeed Aldossary

Email: [msfd99@hotmail.com](mailto:msfd99@hotmail.com)

**Abstract:** This case describes the management of a 12 year-old boy with mild hemophilia A, who was referred to Department of Pediatric Dentistry for orthodontic extractions. This case was treated under Desmopressin and tranexamic acid cover with caution related to local anesthesia administration technique which highlighted the importance of close and detailed liaison with the hematology team to formulate a comprehensive treatment plan.

**Keywords:** Hemophilia A; Bleeding disorder; DDAVP; Tranexamic acid

### INTRODUCTION

Hemophilia A is an X-linked recessive deficiency in factor VIII and is the commonest of all types of hemophilia. The degree of severity is varied: severe (<1 IU/dl of factor VIII), moderate (1-5 IU/dl), mild (6-40 IU/dl) and carriers who are treated as mild hemophiliacs if the factor level is <50 IU/dl [1]. The normal range of factor VIII is 50-100 IU/dl. Hemophilia A affects males and is characterized by increased bleeding tendency. Treatment modalities include: factor replacement, Desmopressin (desamino-8-D-arginine vasopressin [DDAVP]) and antifibrinolytic agents (tranexamic acid) [2].

For dental management of patients with hemophilia, it is important to know the child's type of hemophilia, the severity and their normal form of medical treatment for controlling bleeding. It is mandatory to be aware of the importance of close and detailed liaison with the hematology team to formulate a comprehensive treatment plan. Most importantly, inform the hematology team of the detailed planned dental treatment and confirm the management plan with them. This case describes the management of a 12 year-old boy with mild hemophilia A, who was referred to Department of Pediatric Dentistry for orthodontic extractions.

### CASE REPORT

TK, a 12 years old male child, was referred from Orthodontic Department to Pediatric Dental Department for extraction of teeth 55, 14, 24, and 65 prior to the provision of fixed orthodontic appliance to correct his protruded maxillary anterior teeth after

failure of functional appliance. The relevant medical history:

- Mild hemophilia A, diagnosed at age of 7 years.
- Baseline level factor VIII = 0.14 IU/ml (under 20%)
- Mild well-controlled asthma.
- Allergy to fish, egg, cheese, nuts and kiwi fruit
- Febrile convulsions as a child
- Jaundice at birth

TK had general anesthesia (GA) at age of 2 years for dental extractions. There was a history, five years ago, of restorations and extractions of primary teeth done under local anesthesia (infiltration, not inferior dental nerve block) in the Dental Unit in the Children's Hospital with DDAVP and tranexamic acid cover. TK showed well response to DDAVP.

TK had an orthodontic functional appliance for 16 months to decrease the overjet (non-extraction) but discontinued because of poor compliance, multiple appliance fractures and poor results.

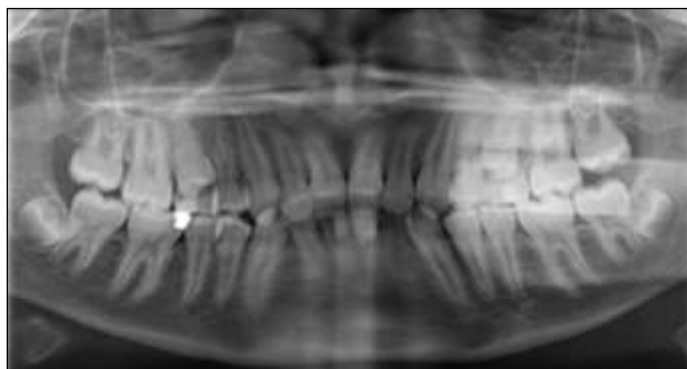
Clinical and radiographic examinations (Figures 1, 2) reveal incompetent lips, fair oral hygiene, partially erupted tooth 27, deep stained fissures and deficient fissure sealant on posterior teeth, recurrent caries under the mesial amalgam restoration in tooth 46, mild generalized diffused enamel opacity, advanced physiological root resorption of teeth 55 and 65, and missing teeth 18 and 28.

Occlusion assessment showed class II division 1 incisor malocclusion, class I right and left molar

relationship, increased overjet (11-12 mm), and 30% overbite.



**Fig-1: Pre-treatment intra-oral views**



**Fig-2: Pre-treatment OPG radiograph**

The aims and objectives of treatment were to improve oral hygiene and reduce caries risk, to facilitate orthodontic treatment to reduce overjet for esthetic reasons and reduce risk of trauma, and to provide safe dental procedures in light of the medical impairment.

The treatment plan included:

**A. Initial management:**

- a. Preventive advice: enhanced prevention including Oral Hygiene Instruction (OHI), dietary advice, topical fluoride varnish and high fluoride toothpaste.
- b. Liaise with hematology team at Children's Hospital to confirm cover requirements and plan dental treatment.

**B. Intermediate management:**

- a. Fissure sealants: molars and premolars.
- b. Restoration of tooth 46.
- c. Extraction of teeth 14, 24, 55, 65.

**C. Long term management:**

- a. Liaise with Orthodontic Department on completion of treatment to confirm start of fixed appliance treatment.
- b. Liaise with General Dental Practitioner for ongoing dental care.
- c. Review and monitor oral hygiene.

The initial assessment visit included clinical and radiographic examination, treatment planning and prevention part included OHI, dietary advice, prescription of 2800 ppm fluoride toothpaste, and application of topical fluoride varnish (2.26%; 22,600 ppm F) (Duraphat® Colgate, Guildford, UK).

I liaised with the hematology team with the treatment plan and later on, a reply from them was received, and they advised:

- a. DDAVP prior to extractions with factor VIII levels measured one hour afterwards to ensure adequate response.
- b. Extractions to be carried out in the Dental Unit in the Children's Hospital.
- c. Restoration to be carried out with infiltration under tranexamic acid cover.

Few weeks later, teeth 55 and 65 exfoliated. Fissure sealants (Delton® Opaque, Dentsply, York, PA, USA) on molars and premolars under cotton wool roll isolation were completed.

In the following dental visit within the pediatric dental department, TK had taken oral (syrup) tranexamic acid dose (25 mg/kg) both morning and afternoon that day.

Under local anesthesia; 4% Articaine with adrenaline 1:100,000 (2.2 ml) (Septanest, Septodont, Saint-Maur-des-Fosses, France) buccal and intrapapillary infiltration, latex-free rubber dam isolation, with clamp 14A on tooth 46, the mesial amalgam restoration was removed followed by complete caries removal. Squiveland matrix was placed and light-cured resin modified glass ionomer liner/base (Vitrebond™, 3M™ ESPE™, St. Paul, MN, USA) and composite restoration (Filtek™ P60, 3M™ ESPE™, St. Paul, MN, USA) were placed in addition to occlusal fissure sealant.

One month later, TK attended early morning to Day Care Unit in the Children's Hospital. I liaised with the hematology team to ensure cover was given in timeous fashion:

- a. IV DDAVP infusion. Factor VIII blood test was done 1 hour post-administration of DDAVP
- b. Hematology team confirmed factor VIII level as 50% and therefore adequate for extractions.

TK also took oral (syrup) tranexamic acid (25 mg/kg), three times a day, in the morning, and this was continued for five days.

In the Dental Unit within the Hospital, under local anesthesia; buccal infiltration of Lidocaine Hydrochloride 2% with 1:80,000 adrenaline (epinephrine) injection solution [Lignospan special, Septodont, Saint-Maur-des-Fosses, France], extraction of teeth 14, 24 was completed. Hemostasis was checked and TK returned to the Day Care Unit and monitored by the hematology team for one hour post-extraction.

Few weeks later, in joint clinic with Orthodontic Department, there was no symptoms, no complaints post-extraction and with good healing of extraction sites (Figures 3).

Records for orthodontics were done by Orthodontic Department and orthodontic bond-up started (Figure 4).



**Fig-3: Intra-oral views pre-fixed appliance bonding**



**Fig-4: Intra-oral view: post-fixed appliance bonding**

The long-term treatment plan and future considerations include:

- a. Reinforce prevention (enhanced prevention).

- b. Complete the fixed orthodontic treatment.
- c. Retention of upper and lower arches post-orthodontic treatment.

d. Liaise with General Dental Practitioner for continued dental care.

## DISCUSSION

TK parents completed our standard medical history proforma. They provided all the information relevant to his medical history and when this information was compared with that in TK's Children's Hospital medical notes prior to treatment, a high level of accuracy was seen. It was however been found that parents/guardians are not always able to accurately report medical history information, particularly in cases of children with a medical condition, necessitating close questioning and review of any information provided prior to treatment by the clinician [3].

It is critical that there is close liaison with the hematologist to ensure appropriate management [1]. This was undertaken highlighted the importance of close and detailed liaison with both the hematology team and the Orthodontic Department to formulate a comprehensive treatment plan.

Although TK was referred for orthodontic extractions, it was essential that a full assessment was undertaken, and an appropriate treatment plan and dental visits arranged carefully to reduce the exposure to prophylactic cover [4].

TK required one restoration and the extraction of two teeth prior to the commencement of orthodontic treatment. As part of his continuing dental care he will require the reinforcement of preventive advice and both clinical and radiographic review.

Patients with mild hemophilia A need pre-operative cover for invasive procedures where bleeding is likely. This will involve the release of endogenous factor stores using DDAVP (if responsive) or direct factor VIII replacement therapy. The aim of the hematological cover was to raise the factor levels to approximately the normal level and maintain them sufficiently long enough to enable a stable blood clot to form [4]. TK had had demonstrated a good response to DDAVP in 2008.

Antifibrinolytics, such as tranexamic acid, should be given orally starting on the day of extraction at a dose of 25 mg/kg every 8 hours for 5-7 days. These agents reduce the need for repeated dose of factor replacement or DDAVP as they minimize the likelihood of delayed post-operative bleeding. There is evidence that using combination of antifibrinolytic therapy and factor replacement or DDAVP, as prescribed in this case, is more effective than using one therapy alone [1].

Recently, there have been reports that the use of Articaine with 1:100,000 epinephrine may achieve more optimal bone penetration. This local anesthetic has been described for infiltration as an alternative to

inferior dental blocks in the restoration of mandibular molars, removing the need for pre-operative factor cover [5].

In addition to hemostatic cover, local hemostatic measures are very important to minimize bleeding [4]. In this case, the rubber dam clamp was placed carefully to minimize damage to gingivae and the extractions of 14 and 24 were carried out atraumatically. Hemostasis was checked while TK was still in the dental clinic and then he was monitored by the hematology team for one hour post-extraction. Other local measures such as suturing and placement of hemostatic agents were not required.

TK was considered to be high caries risk because of his medical condition, in addition to presence of plaque deposits, high sugar intake and the presence of the fixed appliance which had the potential to increase plaque retention. Enhanced prevention was implemented including OHI, topical fluoride application, dietary advice and prescription of 2800 ppm fluoride toothpaste [6].

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