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CASE REPORT

Hemophilia A. Considerations for dental management of pediatric patients.

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Abstract: A male pediatric (9 years and 10 months old) patient, who had a presumptive diagnosis of hemophilia due to persistent bleeding after treatment with steel crowns made in an earlier appointment, came for consultation to the Faculty of Dentistry at the Universidad Autónoma de Nuevo León. Interconsultation was carried out by a hematologist who noticed coagulation factor VIII decreased through laboratory examinations confirming hemophilia A diagnosis. A complete dental treatment was planned and conducted together with the hematologist who gave directions to prepare the patient with missing clotting factor replacement by cryoprecipitates or with factor VIII concentrate intravenously before and after the dental intervention in the hospital. The aim of the article is to highlight that hemophilia is a disease which can be detected during dental consultation in some patients and, for them to be successfully treated, a multidisciplinary management protocol involving hematologists and dentists is required.

Keywords: *Treatment, patient, child, hemophilia A.*

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INTRODUCTION.

Hemostasis is a defense mechanism which protects the body from losing blood after a vascular injury. It has been clasified into: primary hemostasis, involving platelet, and coagulation phase or secondary hemostasis[1,2].

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. Their frequencies are 1 in 5,000 and 1 in 30,000 male alive births, respectively[3].

Men have only one X chromosome, so if the factor VIII gene on that chromosome is defective, they will have hemophilia A. Thus, most people with hemophilia A are males (Figure 1).

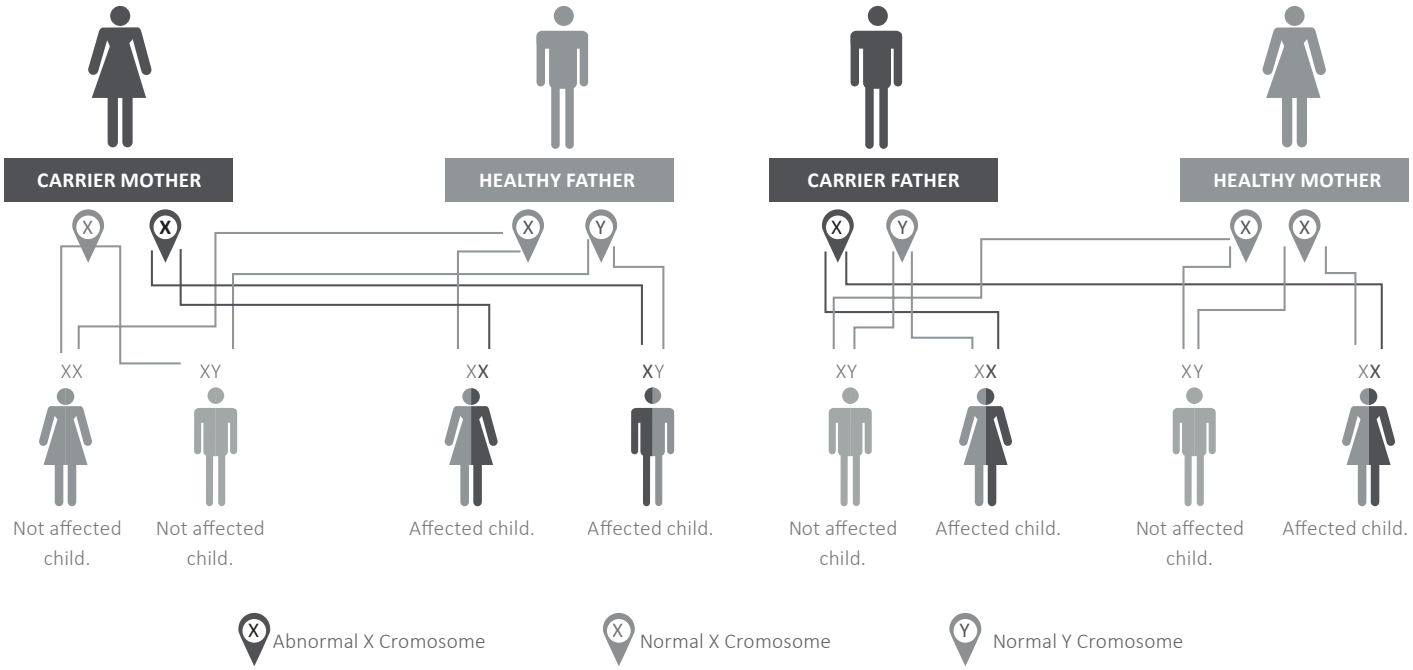
As a monogenic disease, it is considered to be ideal for promising treatment with gene therapy during the last decade[4,5].

Its manifestations include hemarthrosis, bleeding into joints and localized or disseminated hematomas. 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% (Figure 2). 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[6-8].

In severe cases, there can be spontaneous or prolonged bleeding without cause during injury, tooth extractions and surgery[9,10].

Diagnosis is made through clinical and family history; however, the disease can be discovered during a dental treatment if there is prolonged bleeding. The definitive diagnosis of suspected hemophilia must be done through laboratory tests to measure clotting factor levels. Among the required exams, there is a flow cytometry, bleeding time (BT), activated partial thromboplastin time (APTT),

Figure 1. Inherited hemophilia patterns.



Source: World Federation of Hemophilia 2008.

Table 2. Classification of disease severity.

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.

prothrombin time (PT) and thrombin time (TT). All results should be normal except for APTT which will show an extension of more than two standard deviations from the mean. Besides, platelet count, clot retraction, platelet aggregation and factor VIII and IX quantification to confirm the diagnosis should be done[6-11].

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[12-14].

Sometimes, a person with hemophilia A or B can create inhibitors directed against factor VIII or IX, after treatment for administrating the missing factor. The antibody attaches to factor VIII or IX and neutralizes its capacity to stop bleeding[15].

On average, about seven thousand Mexicans have hemophilia and about a third does not know they have it. This is relevant because of the risks involved in dental procedures and the possible post operative complications[6]. In this article, the case report of a patient with hemophilia is shown and the main aim is to assess considerations for its management.

CASE.

The male 9 years and 10 months old patient consulted the Graduate Odontopediatrics Department at the Faculty of Dentistry of the Universidad Autónoma de Nuevo León for presenting bleeding after treatment with stainless steel crowns. Upon interrogating his mother for medical history, family history for hemophilia was negative. According to dental history, apexification treatment had been performed in tooth 46 and restorations in upper and lower right quadrants

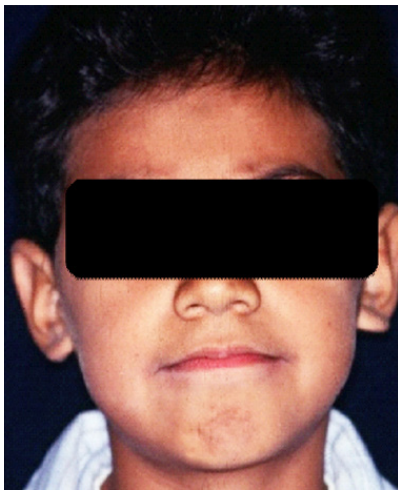


Figure 3. Extraoral examination front view.

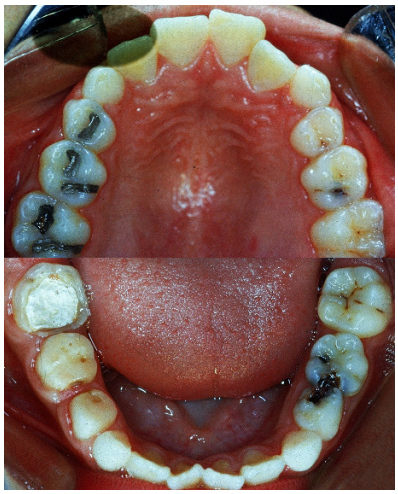


Figure 4. Intraoral examination.

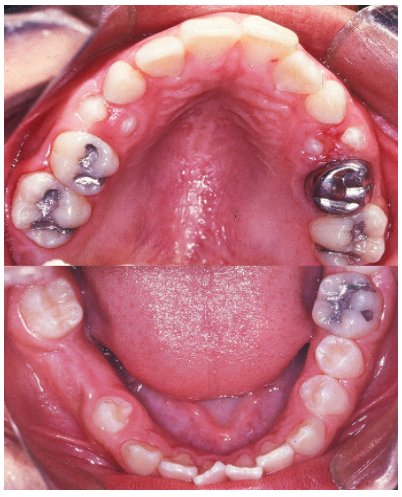


Figure 5. Treatment and check-up at six months.

included two steel crowns, but because of a subsequent abnormal bleeding, steel crowns were removed and he was referred to the graduate department.

In interconsultation, a pediatrician and a hematologist, who noticed coagulation factor VIII decreased and APTT increased through laboratory examinations, confirmed the diagnosis of moderate hemophilia A.

In the extra oral examination, the patient's skin and tissues had their natural colors (Figure 3). In the intraoral examination, he showed generalized swollen gum due to poor oral hygiene, early mixed dentition, amalgam restorations in teeth 16, 55 and 54; incisal fracture in 21, moderate caries in 64, 65, 26 and 36; deep caries in 75 and 74; preparations for crown in 84 and 85; apexification in 46, and teeth in wrong position (Figure 4). The diagnosis was confirmed by ancillary diagnostic tests, photographs and radiographs.

TREATMENT.

A complete dental treatment was planned and conducted together with the hematologist who gave directions to prepare the patient with missing clotting factor replacement by cryoprecipitates or with factor VIII concentrate intravenously before and after the dental intervention to prevent postoperative complications in the hospital. For each stage, 700U of factor VIII were administered. Treatment was done in two appointments or stages as follows: on the first appointment, left quadrants were restored prior hematological management, putting local anesthesia and removing calculus, performing restorations and extractions. After a week, control photographs were reviewed and they showed good evolution of the healing process.

In interconsultation, an endodontist and orthodontist examined teeth 46, which presented apexification and radiographical injury in the furcation area and, because of the prognosis, extraction and posterior orthodontic treatment evaluation was indicated.

During the second stage and before hospital preparation by the hematologist, local anesthesia was put and restorations and extractions were performed in right quadrants. Also, in the follow-up appointment a week after,

a good evolution of the healing process was observed.

Clinical examinations with photographs and radiographs at two weeks, three and six months to assess tooth eruption process were indicated (Figure 5).

Additionally, clinical and radiographic examinations every six months and control of dental plaque and fluoride application every three months was recommended.

DISCUSSION.

The protocol for managing patients with hematological disorders is paramount. According to the Clinical Practice Guideline for Diagnosis and Treatment of Pediatric Hemophilia in Mexico described by the Ministry of Health in 2009 and updated in 2012, it is recommended that all patients with hemophilia in the country are treated with recombinant or plasma-derived clotting factor concentrates and treatment success lies mostly in multidisciplinary management especially between hematologists and dentists[6,16,17].

It is important to take into account these guidelines for management and dental treatment of these patients. The dose for factor VIII is calculated by taking the patient's weight in kilograms and multiplying the factor level desired times 0.5. The result will indicate the number of factor units required[6]. 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. Generally, the treatment of mandibular molars is performed after blocking the alveolar nerve. This type of anesthesia can only be applied after increasing coagulation factor levels to 50% using an appropriate replacement therapy because of muscular bleeding risk, together with likely involvement of airways due to hematoma formation in the retromolar or pterygoid space. It is always advisable to use absolute isolation in care to prevent accidents and anesthetic with vasoconstrictor to produce additional local hemostasis[18].

For dental management, professionals should understand the disease and its complications and extensively explain the possible implications in cases of invasive treatments to patients with bleeding risk[19].

CONCLUSION.

Patients with bleeding disorders are considered high-risk to be treated in the dental office due to their severity and complications.

Not all patients know about their disease, so it is important for the dentist to consider diagnosing hemophilia during consultation. It is essential to know and take into account the clinical practice guidelines and work in multidisciplinary teams including hematologists to establish a correct diagnosis and treatment plan[6,16-18].

We emphasize the importance for hemophilia patients to receive the fewest treatments. Therefore, prevention becomes critical for them with regular check-ups, dental education for children and their parents, fluoride applications, early placement of sealants and eruption control and coordinated comprehensive multidisciplinary care by a professional health team, according to the protocols accepted in use and, if any, to the national treatment guidelines. These are essential considerations for management which contributes significantly to maintain oral health in these patients.

Hemofilia tipo A. Consideraciones en el manejo odontológico de pacientes infantiles.

Resumen: Se presenta a consulta en la Facultad de Odontología de la Universidad Autónoma de Nuevo León paciente masculino pediátrico de 9 años 10 meses, el cual ingresa con un diagnóstico de presunción de hemofilia debido a un sangrado persistente posterior al tratamiento con coronas de acero realizadas en una cita anterior. Se efectúa interconsulta con el hematólogo quien mediante exámenes de laboratorio observa una disminución del factor VIII de coagulación lo que confirma el diagnóstico de hemofilia tipo A. Se planea y realiza el tratamiento integral odontológico

en equipo con el hematólogo quien indica que se prepare al paciente a nivel hospitalario con la reposición del factor de coagulación faltante a través de crio precipitados o mediante concentrado del factor VIII por vía intravenosa previo y posterior a su intervención dental. El objetivo del artículo es destacar que la hemofilia puede ser una enfermedad detectada durante la consulta dental en algunos pacientes y que para que éstos sean tratados con éxito se requiere un protocolo del manejo multidisciplinario entre hematólogos y odontólogos.

Palabras clave: *Tratamiento, paciente, infantil, hemofilia A.*

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