EVALUATION OF THE ORAL AND DENTAL HEALTH OF CHILDREN WITH HEMOPHILIA IN JORDAN
EVALUATION OF THE ORAL AND DENTAL HEALTH OF CHILDREN WITH HEMOPHILIA IN JORDAN

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EVALUATION OF THE ORAL AND DENTAL HEALTH OF CHILDREN WITH HEMOPHILIA IN JORDAN

By
Iman Said Nazzal

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July, 2013
نحن الموقعين أدناه، نتعهد بمنح جامعة العلوم والتكنولوجيا الأردنية حرية التصرف في نشر محتوى الرسالة الجامعية، بحيث تعود حقوق الملكية الفكرية لرسالة الماجستير إلى الجامعة وفق القوانين والأنظمة والتعليمات المتعلقة بالملكية الفكرية وبراءة الاختراع.

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DEDICATION

Thank you Allah for giving me the strength to reach this day

I dedicate this work to….

The four pillars of my life: My husband, my parents, my parents in law and my little angel Nour.

Without you, my life would fall apart. I might not know where the life’s road will take me, but walking with You, God, through this journey has given me strength.

Mohannad, you are everything for me, without your love and understanding I would not be able to make it.

Mom, you have given me so much, thanks for your faith in me, and for teaching me that I should never surrender.

Daddy, you always told me to “reach for the stars.” I think I got my first one. Thanks for inspiring my love for transportation.

We made it…
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Furthermore I would also like to acknowledge with much appreciation the crucial role of my thesis committee Dr. Suhad Al-Jundi and Dr. Fuad Mhairat who have invested their effort in guiding me to achieve the goal.

A special thanks goes to my class mates, Abeer Al-Batta, Huda Mashal, and Wasan Khawaja who helped me to assemble the parts and gave suggestion about the task. Last but not least, many thanks go to Dr. Awidi Al-Abbadi and Dr. Mousa Barqawi, who gave me the permission to use all required data and the necessary information to complete the task.
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ABSTRACT

EVALUATION OF THE ORAL AND DENTAL HEALTH OF CHILDREN WITH HEMOPHILIA IN JORDAN

By
Iman Said Nazzal

Introduction: Hemophilia is considered one of the most common bleeding diathesis in children, oral health of patients with hemophilia is not important in developing region, because they usually get less oral health treatment than the healthy population; however they may have complications that can interfere with their health.

Aims: The aim of this study is to measure the prevalence of dental caries, gingival plaque, and gingivitis condition in children having hemophilia and match it to healthy children in a sample of Jordanian children.

Methods and Materials: This is a case-control study from north and middle districts in Jordan, twenty six children affected with hemophilia were recruited from several hospitals in north and middle districts in Jordan, and they were matched in sex and age with healthy unaffected children. Indices were registered for decayed, missing, and filled surfaces and teeth in both dentitions (dmfs/dmft) / (DMFS/DMFT). The plaque and gingivitis scores were also measured.

Results: children with hemophilia had lower bacterial plaque scores than healthy children in permanent dentition but slightly higher scores in primary dentition. The gingival scores were also lower in children with hemophilia in both primary and permanent dentition. For dental caries scores, the DMFS/DMFT scores were higher in children with hemophilia but the difference was not significantly different. However the dmfs/dmft scores were higher in healthy children, but further analysis showed the difference was due to significantly higher scores of filled surfaces and higher scores of filled teeth.

Conclusion: Dental care for children with hemophilia in most developing countries including Jordan is minimal, precise dental prevention programs can greatly reduce the need for restorative treatment and avoid the need to extract teeth.
Chapter One: Literature Review

1.1. Hemostasis

The known method for blood coagulation is a complicated chain of steps composing from the interaction of the bleeding blood vessel, platelets, with over 20 variable kinds of proteins [Aster & Robbins, 2003].

When a blood vessel bleeds due to an injury, platelets gather near the affected area and shape a temporary clot to stop more bleeding, which is very disorganized to act as a long-term plug, so a cosequenced chemical steps occur, leading to the formation of a more dependable clot. Fibrin producing needs the interaction of multiple chemicals, in particular a cascade clotting factors which are thirteen variant clotting factors are recognized [Aster & Robbins, 2003].

The clotting cascade can be defined as the sequence of steps needed to organize end plug of fibrin. The cascade amplifies to produce a correct size from fibrin initially started to be activated by the insult.

Certain clotting factors in hemophilia disease are either deficient, improperly formed or absent. The clotting pathway amplifies to effectively block up the injured location, deficiency or breakdown of one clotting factor only can extremely prolong bleeding time [Cawson & Scully, 2000].
1.2. Hemophilia

1.2.1 Definition and Prevalence

Hemophilia, a deficiency of clotting factors VIII or IX, is a rare, sex "X-linked" congenital disorder that results in absence of physiological hemostasis and appeared with spontaneous or prolonged bleeding. Repetitive episodes, usually in the joints and muscles can also lead to long-term disability [Aster & Robbins, 2003].

Hemophilia is considered one of the most commonly bleeding diathesis in children [Saxena and shashikiran, 2010]. It is thought to be the most common sex-linked bleeding disorder worldwide, with an incidence of five thousands to ten thousands male births [Kabil et al., 2007; Israels et al., 2006].

1.2.1.1 Inheritance patterns

Hemophilia A and B are sex "X- linked" recessive inherited disease caused by mutations on the factor VIII and factor IX genes on the X chromosome [Aster & Robbins, 2003]. The variance in the size of the genes explains the variance in prevalence between the two hemophilia types; the bigger size of factor VIII gene, gives the mutation higher possibility to occur [Israels et al., 2006]. Defects in the Factor IX gene may also be due to either large deletions or point mutations. Patients with large deletions and absent protein may be at risk of anaphylaxis when treated with Factor IX replacement [Israels et al., 2006].

Interestingly, females carrying the mutation do not bleed spontaneously, but they tend to exhibit less degrees of clotting factor and can be susceptible to severe bleeding after being traumatized [Stachnik, 2010].
1.2.2. Classifications of Hemophilia

Hemophilia is usually classified in regard to factors deficient. Hemophilia A or named "classic hemophilia" is when Factor VIII is deficient which happens in 85% of patients [WFH, 2005]. Hemophilia B or Christmas disease is a deficiency of Factor IX, in 10-15% of hemophilia cases [WFH, 2005].

Some reports considered a third type of hemophilia; Hemophilia C, and defined it as deficiency of Plasma thromboplastin antecedent (PTA) [Saxena and shashikiran, 2010].

Von Willebrand disease is characterized by a reduced quantity of circulating vWillebrand factor (vWF); which stabilizes factor VIII by binding to it. A deficiency in vWF is associated with a secondary decrease in the levels of factor VIII. There are other less common variants of this disease including qualitative and quantitative defects in vWF [Aster & Robbins, 2003].

Further classification of patients with hemophilia can describe the quantity of factor VIII or IX. This classification is based on blood levels of Factor VIII and Factor IX as shown in Table (1.1).

<table>
<thead>
<tr>
<th>Severity</th>
<th>Clotting factor level % activity (IU/mL)</th>
<th>Bleeding episode</th>
</tr>
</thead>
<tbody>
<tr>
<td>Severe</td>
<td>1% (&lt; 0.01)</td>
<td>Spontaneous bleeding, mainly in joints and muscles</td>
</tr>
<tr>
<td>Moderate</td>
<td>1%-5% (0.01-0.05)</td>
<td>Spontaneous bleeding occasionally. Profused bleeds following accidents or surgery</td>
</tr>
<tr>
<td>Mild</td>
<td>6%-40% (0.06-0.40)</td>
<td>Profused bleeding after large trauma or surgery</td>
</tr>
</tbody>
</table>
Patients with *severe hemophilia* have <0.01 IU mL Factor VIII or Factor IX activity. Cases with the inherited hemophilia, two-thirds of them exhibit severe disease [Davis et al., 2009].

In contrast, patients with *mild hemophilia*, having 0.06–0.40 IU residual Factor VIII or Factor IX, do not usually bleed spontaneously [Stachnik, 2010]. Bleeding episodes in patients with mild hemophilia happen only after being traumatized or after surgical intervention. Usually, patients with mild hemophilia have no limited functions. Interestingly, carriers can be treated like mild hemophilia if their Factor VIII level is less than 0.50 IU/dl [Brewer et al., 2003].

*Moderate hemophilia*, defined as 0.01–0.05 IU residual Factor VIII or Factor IX activity, is diagnosed in 15–20% of patients with hemophilia. It can be predicted that bleeding quantity, joint motion and complications in daily life of patients with moderate hemophilia are in the middle of those with severe and mild hemophilia. Treatment of bleeding episodes in moderate hemophilia is often postponed, may because of little home care and because of more difficult way to detect bleed [WFH, 2005]. This may lead to under-estimation of treatment for cases with moderate hemophilia [Den Uilj et al., 2009]. The median age to diagnose a patient with severe hemophilia is in average at 1 month, while in patients have moderate deficiency their condition usually diagnosed during the first years of life but mild hemophilia may go undetected or undiagnosed for years [Stachnik, 2010].

### 1.2.3. Clinical Implications

Bleeding accidents noticed in individuals with hemophilia differs with age. Patients with hemophilia often usually show the disease during neonatal life with profused bleeding from the umbilical cord or intracranial bleeding [Davis et al., 2009]. Children
ages between "1 and 6 months" may be prone to bleeding from soft tissues. Toddlers also can demonstrate bleeding from the gingiva during teething [Davis et al., 2009]. Because child motion is increased with age, the joint might bleed more oftenly. In older ages, the joints bleeds most commonly which can lead to "hemarthrosis" and compose for seventy to eighty percent of bleeding episodes [WFH, 2005].

In the literature there are some case reports on individuals presenting with multiple hemorrhagic subcutaneous nodules as a first symptom of hemophilia. This sign must give an alarm to the dermatologist to check a hemorrhagic disorder in those cases [Ljung et al., 1990; Davis et al., 2009].

In Ljung et al study to decide the most commonly showing sign in patients having severe and moderate hemophilia A and B. They reported the mostly seen symptoms to be subcutaneous bleedings in 48% of those severely affected, with a mean age of presentation of 9 months [Ljung et al., 1990].

Bleeding occurs most frequently in patients with hemophilia in the musculoskeletal system; it is also counted as the major source of morbidity in individuals with insufficient treatment [Brown, 2005]. The World Federation of Hemophilia classified the site of bleeding into two main categories; serious sites and life threatening sites. Serious sites are: Joints (hemarthrosis) with incidence of 70-80%, muscle (soft tissue) with incidence of 10-20%, mouth/ gum/ nose and hematuria. Life threatening sites are: CNS with incidence of <5%, gastrointestinal, neck/throat, and severe trauma [WFH, 2005].

In developing countries, most of the patients with hemophilia are in the pediatric age group as they seldom reach adulthood because of inadequate treatment [Chuansumrit, 2003].
The dentist in many situations can be the first to discover the disorder in a patient [Kumar et al., 2007]. The oral cavity with its rich vascularity is surly a high area for hemorrhage or excessive bleeding in this group of patients. Unfortunately, many hemophiliacs tend to neglect oral and dental health for fear of bleeding during dental treatment procedures and this may be the primary reason for absence of effective dental care for them [Kumar et al., 2007]. Mild hemophilia may be still undiagnosed until adolescence or may be later especially if the individual was not prone to a major surgery, severely traumatized or have teeth extracted during his life. Frenum and tongue are the most commonly recognized site of oral bleeding [Kumar et al., 2007].

1.2.4. Long Term Complications

A well known disability of hemophilia is permanent destruction to the joints (hemarthropathy) caused by repetitive episodes of bleeding [Stachnik, 2010]. Chronic changes occur in a joint affected with recurrent bleeding episodes (target joint). [Stachnik, 2010]. Repetitive muscular bleeding accidents can also result in long-term disability like contractures [Stachnik. 2010].

If a specific joint had repetitive bleeding episodes in the synovium appears inflamed in a chronic way and finally get enlarged, causing the affected joint to look obviously enlarged and swollen. This swelling is usually painless; hence, joint movement can be preserved to some extent. WFH advised daily exercises to strengthen muscles and improve or maintain joint mobility. Sufficient levels and frequency of factor concentrate supplements should be considered to avoid frequent bleeding. Even though, when factor is present in enough levels, short treatment periods (6-8 weeks) of prophylaxis in combination with physiotherapy is satisfactory joined with intra-articular injection of a long-acting steroid [WFH, 2005].
Muscle bleeding and hemarthrosis are most commonly seen in chronic features in hemophilic children [Davis et al., 2009; Stachnik, 2010].

1.2.5. Hematology Laboratory Tests

Prothrombin time (PT) and activated partial thromboplastin time (PTT) are coagulation screening tests performed using citrated plasma. The PT is prolonged in deficiencies of fibrinogen, prothrombin, and Factors V, VII, or X. Also congenital factor deficiencies, and other conditions prolong readings of PT [Stachnik, 2010].

The activated partial thromboplastin time (PTT) measures the contact system and the intrinsic and common pathways. The PTT is prolonged when Factors II, V, VIII, IX, X, XI, XII are deficient. Bleeding time estimates platelets functions (Quality) and platelet count estimates platelet quantity [WFD, 2005].

Screening tests result will be a prolonged (PTT) in severe and moderate hemophilia. An accurate diagnosis relies on factor assay to show deficiency of Factor VIII or Factor IX [WFD, 2005; Stachnik, 2010]. The laboratory tests and their outcome in hemophilia are shown in Table 1.2.

Table 1.2: Laboratory Tests' Results in Hemophilia [Adapted from Stachnik, 2010]

<table>
<thead>
<tr>
<th>Test</th>
<th>Specifications</th>
<th>Outcome in Hemophilia</th>
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<tr>
<td>Bleeding time</td>
<td>Platelet</td>
<td>Normal</td>
</tr>
<tr>
<td></td>
<td>Subendothelial interaction</td>
<td></td>
</tr>
<tr>
<td>Platelet count</td>
<td>Platelet per µL</td>
<td>Normal</td>
</tr>
<tr>
<td>PT</td>
<td>Factors I,II, V, VII, X</td>
<td>Normal</td>
</tr>
<tr>
<td>PTT</td>
<td>Factors I , II, V, VIII, IX, X</td>
<td>Prolonged</td>
</tr>
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Prior to laboratory testing patients should be instructed to fast before performing blood tests. Excess lipids will be cleared from the blood after fasting; which when not cleared may alter the proteins quantity in automatic analyzers. Patients should be avoided to take drugs which will alter the final results; ASA-acetylsalicylic acid, is an example which can dramatically affect platelets work and falsely give elongated bleeding time [WFH, 2005].

1.2.6. Treatment of Hemophilia

The main goal of treatment is to prevent or stop bleeding episodes. Both non-pharmacologic adjunctive therapy and pharmacologic therapy are ways for the management of this disease [Stachnik. 2010].

1.2.6.1. Non-pharmacologic Therapy

Non-pharmacologic therapy is defined as supportive therapy to control bleeding episodes in joints or muscles [WFH, 2005]. Splints, crutches, casts, or even wheelchairs are recommended to help the affected muscle or joint to rest following a bleeding episode. Ice packs must be used over the affected area for twenty minutes every four to six hours to reduce the inflammation. When swelling is healed, physiotherapy should start to keep joint and muscle function [Stachnik, 2010].

1.2.6.2. Pharmacologic therapy

Desmopressin Acetate

It is a synthetic analogue of the naturally occurring hormone arginine vasopressin and can be used in the treatment of patients diagnosed with mild hemophilia A and some cases of moderate hemophilia A [Franchini & Lippi, 2011].
However, the literature has reported that some patients who were given desmopressin very frequently showed tachyphylaxis "i.e. progressively poor response" [Castaman, 2008; Franchini & Lippi, 2011].

Following the administration of desmopressin, blood plasma Factor VIII levels elevate two to six folds then it returns back to baseline level within six to fourteen hours. The elevation in Factor VIII levels clarifies the truth that patients affected severe hemophilia A are not eligible for this kind of treatment [Ozgonenel et al., 2007].

Instructions should be given to patients to reduce water ingestion during desmopressin treatment and to stop the treatment using desmopressin after three successive daily doses to minimize the risk of hyponatraemia—which is defined as an electrolyte disturbance in which the sodium ion concentration in the serum is lower than normal [Wong & Recht, 2011]. Other untoward complications are in general mild and include headache, nausea, facial flushing, and increased heart rate or blood pressure [Wong & Recht, 2011].

After all, the effects of desmopressin are temporal but repetitive doses can lead to tachyphylaxis. Therefore, the advantages of desmopressin are mainly fro those individuals who needs high factor degrees for extended periods [Stachnik, 2010].

In addition, it has been reported that hemophilic children may have a lower rate of biologic response compared to adults and a minority of adult patients are not able to attain clinically useful Factor VIII levels post-desmopressin administration [Castaman, 2008].

**Antifibrinolytic Agents**

Two antifibrinolytic drugs are useful to manage hemophilia diseases in different situations: Tranexamic acid and Aminocaproic acid [Stachnik, 2010]. Because they don't stop a bleeding episode, they are only considered as adjunctive therapy and they are
especially useful for some dental treatments and to stop bleeding from like epistaxis [Gupta et al., 2007].

Many clinical trials over several decades proved that regular single treatment with tranexamic acid alone has modest effect in prevention of hemarthroses in hemophilic patients [Hvas et al., 2007]. However, it is known that it has a value in stopping bleeding from mucosal surfaces in hemophilia and is mainly helpful in dental surgery settings, and in controlling oral bleeding during physiological eruption of teeth [WFH, 2005; Gupta et al., 2007].

**Factor Concentrates**

The first line treatment is the deficient factor replacement with a factor concentrate for both types of hemophilia A and B. Some patients with mild hemophilia A can be treated efficiently only by desmopressin therapy provided that the episode is not severe or life-threatening, but in the case of moderate or severe hemophilia, factor replacement therapy cannot be replaced with treatment methods [Stachnik, 2010].

The aim of replacement therapy is to increase the blood plasma factor degrees to a satisfactory level to get good hemostasis [Stachnik, 2010].

The factor concentrates can be delivered to the hemophilic patients in two ways; intermittent bolus injections or continuous infusion. When comparing intermittent bolus injections of Factor VIII with continuous infusion; there are less clearance, decreased factor usage, and lower treatment problems with the continuous infusion are reported [Martinowitz et al., 2009]. Continuous infusion during surgery has benefits such as providing stable administration of replacement factor to the patient, preventing elevated peaks and low troughs, and moreover, an adequate reduction of factor usage may be achieved by continuous factor infusion [Batorova et al., 2000].
Episodic, or 'on demand' factor replacement therapy is the standard treatment method in which the deficient factor is replaced once the bleeding symptoms are started or as soon as possible [Martinowitz et al., 2009]. More recently, prophylactic administration of clotting factor given to avoid bleeding, is highly recommended as the golden treatment by the Medical and Scientific Advisory Council of the National Hemophilia Foundation, the World Federation of Hemophilia and the WHO [WFH, 2005; NHF, 2013].

Realizing that patients with mild hemophilia rarely expressed spontaneous bleeding gave the idea of primary prophylaxis which can lead to much better joint function and motion preservation [Brown, 2005; Mannucci et al., 2011].

In the mean time, the most famously suggested advised protocol for factor concentrates prophylaxis is the infusion of 25-40 IU/kg in the week 3 times for those with hemophilia A, and twice a week for those with hemophilia B [WFH, 2005]. In patients suffering from repeated bleeding episodes, especially into specific joints, short prophylaxis turns for four to eight weeks is helpful to break the bleeding. This is better to be integrated with synoviorthesis or physiotherapy [Khreisat et al., 2000]. It advisable to administrate a prophylactic dose of clotting factor concentrates before participating in high risk activities to prevent bleeding caused by injury during these activities [Mannucci et al., 2011]. However, it is well known that there is no definite single protocol for prophylaxis even in one country. Currently, different protocols for prophylaxis are being studied and assessed [Feldman et al., 2006; Gingeri et al., 2011]. The most traditional prophylaxis regimes, consists of 2-3 factor infusion per week [Gingeri et al., 2011], however, they are of difficult functionality for many children and their families. A way was developed in Canada to overcome this drawback which applies a single weekly infusion of the deficient factor [Feldman et al., 2006].
However, prophylaxis kept joint function and improves life quality by minimizing joint bleeding episodes [WFH, 2005; Wong & Recht, 2011]. Therefore, in the long term it is considered cost effective since it eliminates and decreases the high cost expressed after morbid joints managing. Cost-efficacy studies aimed to recognize the very minimal requirements are highly recommended so they can minimize the high burden to get treated and facilitate ways to have prophylaxis routinely in most of the world countries [NHF, 2013]. The high cost and repetitive venipuncturing are disadvantages for primary prophylaxis, it not easy to introduce a venous access device in younger children that has to be kept accurately clean and be disinfected to eliminate problems originated from infections and be vigorously cleaned with a water flush following ever factor administration time to avoid the development of clots in the line [Wong & Recht, 2011].

Factor VIII and IX concentrates available in the market are either plasma-derived or generated by recombinant technology [Mannucci, 2011]. To date, it is not clear whether the use of specific Factor VIII product types (i.e. recombinant or plasma-derived Factor VIII products) has an effect on developing inhibitors [Ettinghauzen et al., 2006]. Some comparative studies indicate a slightly lower inhibitor incidence in hemophilia A treated with a unique plasma-derived Factor VIII product [Ettinghauzen et al., 2006].

Two main concerns are associated with factor replacement; transmitting of pathogenic viruses or prions and inhibitors development.

**Viruses’ transmission**

When Hepatitis B and C were discovered in the factor supplements, thoughts were to accept the risk of those infections justified by increased quality of life [Wong & Recht, 2011]. However, after individuals with hemophilia presented with AIDS are discovered, this was received as red alarm, and only then HIV was isolated by heating of factor [Wong & Recht, 2011].
Virus reduction techniques can remove or inactivate any possible viruses originating from the factor original source by treating it by detergent or more efficiently by vapor-heat or pasteurization [Wong & Recht, 2011]. Effectively elimination of viruses by size is achieved by Nanofiltration method. Two virus reduction methods are usually used to increase the efficacy in reducing any potential viruses threat [Stachnik, 2010].

Strongly cleaned forms of Factor VIII concentrates, which was produced by recombinant methods are chosen first because of their better viral safety [Gupta et al., 2007].

**Inhibitor development**

The most critical treatment-related complication in hemophilia today is antibody formation against Factor VIII or Factor IX. In both hemophilia A and B, these antibodies neutralize the action of the replaced clotting factors concentrates. In addition, a severe complication can occur with Factor IX inhibitors; which is anaphylaxis [Wong & Recht, 2005].

If inhibitors are developed, this will not lead to more severe bleeding or more frequent episodes; but they complicate the treatment of a bleeding episode by decreasing the factory efficiency. Inhibitor development is reported in individuals having hemophilia A more frequently than in hemophilia B individuals, and is registered to reach a high number "up to thirty eight percent of patients with hemophilia A" [Stachnik, 2010]. WFH has reported in the published guidelines of year 2005 that about ten to fifteen percent of hemophilia A individuals and only one to three percent of hemophilia B patients are susceptible to manifest with persistent inhibitors making treatments using factor concentrates replacements difficult. While Wong & Recht reported the incidence of this untoward reaction of the immune system is fifteen to fifty percent in hemophilia A and one to three per cent in severe hemophilia B. IgG antibodies, with subtype 4 being the most
predominant inhibitors. In most cases, inhibitors appear after a median of 8-20 exposure days [Wong & Recht, 2005].

Many Factors can increase the chance of inhibitor development: the genetic mutation category, severity of hemophilia, previous history in family, and finally first treatment age. Patients who have severe gene defects such as gene deletion or inversion, frame shift and nonsense mutations are more prone to have inhibitors [Wong & Recht, 2005; WFH, 2005].

FEIBA (factor VIII inhibitor bypassing activity), an activated prothrombin complex concentrate (aPCC), has been recognized as a hemostatic bypassing agent in patients with high-responding inhibitors for decades [Astermark et al., 2012]. More recently, NovoSeven, recombinant Factor VII, has been also known as an effective treatment modality. The variable bypassing mechanisms of these agents are still not fully explained or understood. However, the previously mentioned two methods are considered able to bypass the Factor VIII–dependent step in the coagulation cascade and enhance hemostasis by promoting thrombin generation [Astermark et al., 2012].

**Gene transfer**

New data showed that it can achievable to perform successful genetic transfer for hemophilic patients [Astermark et al., 2012]. The subjective reporting by patients of decreased bleeding episodes strongly highlights that adequate factor levels might be achieved and that a drastic improvement in the treatment of hemophilia is promising [Rick et al., 2003]. Gene transfer for hemophilia requires a combination of animal models, vector delivery systems, and clinical studies aimed to answer specific questions[Rick et al., 2003].
1.2.7. Dental Treatment For Patients With Hemophilia

The most common dental diseases are dental caries and periodontal disease. The management of these conditions for an adult with a congenital bleeding disorder may require the use of clotting factors [Brewer et al., 2003].

Thorough dental and medical testing is mandatory before commencement of treatment, particularly if an invasive dental procedure is in the treatment plan. Medical history and thorough patient assessment should start with standard specified medical questionnaires. Past history of prolonged bleeding surgery or injury should be explained, and patient should be asked if spontaneous bleeding and easy bruising is frequently evident [Gupta et al., 2007].

If the procedure is considered simple and not invasive treatment and the individual manifests mild hemophilia, minimal or no treatment alteration will be needed. The goal of treatment in patients who have severe bleeding disorders, is to decrease the patient challenge by trying to restore the hemostatic system to a satisfactory levels and preserving hemostasis by local and adjunctive ways, keeping in mind that consultation of patient's physician is mandatory before any invasive treatment is performed [Gupta et al., 2007]. In hemophilia, most published guidelines highly recommend the treatment using factor replacement before getting involved into oral surgical intervention [WFH, 2006].

Successful protocols generate from the cooperation between the dentist and the hematologists. Ideally, the dental team which provides dental care for hemophilic patients, must perform an integral part of the overall participating medical team, preferably, the dental team must be located near to the hemophilic center, so as to facilitate access for dental treatment and to encourage both the physician and the patient to get the ideal preventive and curative dental care [Harrington et al., 2000].
The general published protocols recommend factor concentrate usage combined with the local hemostatic measures, like suturs, oxidized cellulose associated with post-operative administration of antifibrinolytic agents [Harrington et al., 2000; Brewer & Correa, 2006]. By using the local measures in some minor oral surgery procedures the use of coagulation factor replacement can be avoided or it can be used only minimally [Brewer & Correa, 2006].

As should be in healthy children, prevention programs are the ideal way to prevent dental emergencies and accompanied complications in hemophilic children. These prevention programs include brushing using fluoride toothpastes, flossing, using of fluoride supplements, sealing the fissures, and controlling the diet of the hemophilic patients [Brewer & Correa, 2006]. Combined preventive methods, can significantly decrease the incidence of dental caries from 70 to 80 % in hemophilic patients [Harington, 2000].

During dental treatment, it is mandatory to prevent any mistaken bleeding episodes from the mucous membranes of the oral cavity while performing a dental treatment in the mouth [Brewer & Correa, 2006]. Injury can be prevented by: careful application of saliva ejectors, careful handling of impressions, placement of X-ray films carefully, especially in the sublingual area; and of course a rubber dam to protect soft tissues during restorative treatment is imperative [Brewer & Correa, 2006].

**Periodontal treatment:** when periodontal tissues are severely diseased, it is advisable to start with supragingival scaling with emphasizing on strict oral hygiene instructions. Subgingival scaling can be performed once the gingival inflammation has reduced [Kumar et al., 2007]. It is a good treatment option to perform the treatment over multiple visits to avoid excessive blood loss [Brewer & Correa, 2006]. Blood loss of during or after
performing periodontal treatment can be managed by local measures using pressure by periodontal dressings in conjunction topical antifibrinolytic agents. However, it should be kept in mind periodontal surgery sometimes can challenge the dentist greater than a simple extraction [Brewer & Correa, 2006].

**Orthodontic treatment:** Orthodontic appliances either removable or fixed ones, are allowed but with standard protocols and hygiene instructions. More strict oral hygiene instructions must be emphasized during treatment of patients with severe hemophilia to ensure that appliances used in orthodontic treatment do not cause damages to the gingiva [Brewer & Correa, 2006].

**Restorative procedures:** Restorative dentistry can be practiced normally make sure that very careful protection of the mucosa is provided [Brewer & Correa, 2006]. A risk of bleeding can be evident matrix bands or wooden wedges are used. Local adjunctive methods or application of topical agents can control bleeding in such cases [Brewer & Correa, 2006]. Harrington recommended that only minimally affected teeth with caries should be restored, and if there is any doubtful prognosis of the restoration to be done it should never be performed in hemophilic patients especially if there is no available factor replacement or it is restricted [Harrington, 2000].

**Endodontics:** Although endodontic treatment is in general considered a low risk reason of bleeding for this group of patients, if a pulpectomy is going to be performed, it is substantial that the step to be done very precisely with the working length [Harrington, 2000; Brewer & Correa, 2006]. Irrigation with sodium hypochlorite in all cases is allowed. Formaldehyde-derived materials can be used when if there is profused bleeding [Brewer & Correa, 2006].
**Dental emergencies:** It is extremely essential to remember that treatment should not be started unless proper planning precedes any procedure as this can lead to additional more serious complications. The most common reported dental complications are pain originated from tooth caries and bleeding from the gingival and periodontal tissues [Brewer & Correa, 2006]. Pain originated from carious teeth is best treated with either pulpectomy or antibiotics so as to give time for the planning correctly of the extraction. Bleeding accompanied from inflamed gingival and periodontal tissues can usually be managed with antibiotics then an appointment with a hygienist can be scheduled [Brewer & Correa, 2006; Kumar et al., 2007].

**Local anesthesia and surgery:** a level of at least thirty percent of normal from clotting factor are needed when infiltration anesthesia is to be performed in the upper or lower jaws and also for periodontal curettage [Brewer & Correa, 2006]. Surgical procedures with more invasive techniques, like inferior nerve block, deep scaling or extraction, standard recommendation is to elevate the factor level 50% of normal pre-operatively, in conjunction with the application of oral antifibrinolytic agent pre- and post-operatively. When the procedures become much more extensive, such as surgical extraction then 100% rise in factor level is necessary in order to eliminate bleeding complications [Harington, 2000; Brewer & Correa, 2006]. It has been reported that 80% chance that hemophilic patients who are not treated with proper doses of prophylactic factor replacement before the commencement of inferior alveolar nerve block injection can have hematomas. Infiltrations, intraosseous, intraligamentary or intrapulpal injections are believed to be safer options [Kumar et al., 2007]. Interestingly, alternative anesthetic techniques like electronic anesthesia deserve to be mentioned in these cases. It is mentioned in the literature to use hypnosis as an alternative pain management technique [Kumar et al., 2007].
Harrington advised performing single extractions in four areas rather than four extractions next to each other, as the latter choice will cause the bleeding site larger and tend to become a large wound that is more difficult to be managed in hemophilic patient [Harrington, 2000].

In some extraction cases where hemophilia is complicated with inhibitor development, some reports suggested the use of Prothrombin complex concentrates or recombinant activated Factor VII, it was proven to be highly effective with no side-effects [Laguna et al., 2005].

Very essential step is to assume that local adjunctive treatment methods to decrease the burden of postoperative bleeding a major treatment modality [Brewer, 2008]. Although several authors recommended that suturing is helpful to avoid postoperative complications of wounds caused by tongue mobility, Brewer reported a minimal number of cases where no suturing was done routinely with any significant elevation in postextraction hemorrhage severity or quantity [Kumar et al., 2007; Brewer, 2008]. However, the pediatric dentist must take the decision based on the age of the patient, number of teeth extracted and severity of the wound [Brewer, 2008].

Local measures aim is to achieve a stable clot within the socket. It usually involves placing a hemostatic agent directly into the area and suturing it in place [Brewer, 2008]. Fibrin glue also has been used with minimal bleeding complications [Brewer, 2008].

**Treatment under general anesthesia:** Factor replacement must be explained with the hematologist before the commencement of treatment day in regard with the dental steps that will be done, preoperative loading dose, and 12-hour after operation a maintenance dose is needed since in most cases the child needs treatment for all of his teeth [Rayen et al., 2011]. Intubation of the airways is risky because mucosal bleeding can happen, avoid nasal intubation because mucosal tissues can bleed [Rayen et al., 2011].
1.2.8. Dental Health in Hemophilic Children

Bad oral health in children affected with chronic illnesses, is considered as a major cause for morbidity and may be classified as risk factor for severe or life threatening situations [Foster et al., 2005].

Children with hemophilia are considered in the High Risk group for dental caries because they show bleeding gingiva, especially following routine tooth brushing and are subjected to chronic periodontal disease [Harington, 2000]. Getting perfect dental and gingival health and dentition free of caries is very important for good child communication, eating, and appearance [Foster et al., 2005]. Chronic infection surrounding some teeth can lead to damage to localized structures, like the successor teeth, further more can force early extraction for primary or permanent teeth, which can cause bleeding complications if the medical condition for those children was not considered and managed in the right way [Brewer, 2008]. Dentists must be perceptible of the effect of bleeding diathesis on the management of patients during dental treatment [Foster et al., 2005]. Moreover, restorative, prophylactic, and surgical dental interventions of patients with bleeding disorders can be best achieved by dentists and practitioners who are aware about the pathology, and complications on dental treatment associated with such conditions [Harrington, 2000; Foster et al., 2005].

1.2.8.1. Prevalence of Dental Caries and Gingivitis in Hemophilic Children

Although many reports dealing with dental management strategies for hemophilics are present, there are only minimal data regarding the dental health for young or old age hemophiliacs [Saxena and shashikiran, 2010]. Recently, new investigations have been published to measure oral health situation in those children.
Many researchers published that caries prevalence in children affected with hemophilia is lower than the healthy population, but these studies were mainly reported in developed countries [Blaszczak, 1999; Sonbol et al., 2001; Zeibolz et al., 2011].

In Poland, prevalence of caries did not differ greatly in children having congenital bleeding diseases compared with healthy children. But on the other hand the dental condition in children having severe hemophilia A and von Willebrand's disease was bad compared to children having other medical problems [Blaszczak, 1999].

In the United Kingdom, Sonbol et al., 2001 concluded that individuals having severe hemophilia have a lower prevalence of dental caries than children who are healthy [Sonbol et al., 2001]. This is mainly because all hemophiliacs in that report visited the dentistry clinic during their reviews to the hematology department and received a better planned prevention program than the normal population.

No clinical difference of oral health (caries and oral hygiene) between patients with congenital coagulation disorders and controls were found in Zeibolz et al., 2011 study in Germany. Furthermore, a strong variance is evident in bone loss of the periodontium, but the registered difference was not clinically meaningful. Unlike previous studies carried out mainly in children, no evidence that dental health or periodontal condition in adult patients with congenital coagulation disorder is worse than that present in healthy subjects in Germany [Zeibolz et al., 2011].

A Turkish study concluded that children affected with hemophilia A have usually significantly higher caries and gingival index scores when comparing those results with the results of matched, healthy control individuals [Baskirt et al., 2009]. In India, it was reported that prevalence of dental caries and treatment requirements was high among hemophilic patients which indicates the need for dental programs at as early as possible for preventive protocols in these patients [Saxena and shashikiran, 2010]. Higher prevalence
of decayed, missed and filled teeth was reported among population suffering from severe type of hemophilia A and B in Pakistan [Azhar et al., 2006]. The results in Kabil et al., 2007 study showed that the DMFT and deft in hemophiliacs were significantly higher than healthy individuals in Egypt, in addition it was higher than those of hemophilic children in developed countries highlighting that the decayed score component demonstrating most of the index score value [Kabil et al., 2007].

1.2.9. Hemophilia in Jordan

Hemophilia A is the most common bleeding disorder in Jordan followed by von Willebrand disease and Glanzmann thrombasthenia [Awidi, 1984; Awidi, 2006]. According to the Jordanian National Registration Record in 2012 only 300 cases were registered as patients with hemophilia, on the other hand more than 200 cases are expected to be present but unregistered. Unfortunately, the data for bleeding disorders originate from cases presented to hospitals or laboratories only, which explains why data about hemophilia are minimal [Awidi et al., 2010].

Although this group of patients has many important and critical considerations regarding their dental health and its direct impact on their quality of life there is no clinical data describing the gingival or dental health in Jordanian children with hemophilia.
Chapter Two: Aims and Objectives

The aim of this work is to describe the oral health condition of children with hemophilia and compare it to healthy children in a sample of Jordanian children, to assess oral diseases' extent in this group of medically affected children.

Since there was no previous clinical researches showed the oral health status and dental caries level of patients with hemophilia, this study may be considered as first attempt to clarify this issue with the following objectives:

1. To investigate the prevalence of dental caries, bacterial dental plaque and gingivitis in children with hemophilia.

2. To establish database on the caries level and oral health status in children with hemophilia, to make data available for future comparisons.
Chapter Three: Methods and Materials

2.1. Study Design

This is a case-control study set to describe the oral health of Jordanian children diagnosed with hemophilia and compare it to healthy Jordanian children. The study was performed in the Northern and Middle of Jordan.

3.2. Ethical Approval

An ethical approval was obtained from the Institutional Review Board (IRB) at Jordan University of Science and Technology [Appendix I]. Proper permissions were obtained from the Ministry of Health and reference hospitals to allow access to examine patients in their facilities [Appendix II, III].

3.3. Patient Recruitment

3.3.1. Study Group

Children having with hemophilia came from hemophilia clinics in Rahma Hospital–Irbid, Al-Basheer Hospital–Amman, Jordan University Hospital–Amman and Royal Medical Services Clinics–Amman.

3.3.2. Control Group
Healthy children were recruited from the Pediatric Dental Clinic in Jordan University for Science and Technology Postgraduate clinics, or from the Orthodontic New Patient Assessment clinic in Jordan University Hospital, they were matched with the
hemophilia group for age and gender. Those were children accompanying patients having dental appointment and not children attending for dental care.

### 3.4. Inclusion Criteria

Children included in this study must be diagnosed with hemophilia type A or B, with a mild, moderate or severe deficiency in the factor level. The age of children participating in this study ranged from three years old to sixteen years old.

### 3.5. Consent

Parents of children of the study and control group were asked to sign an informed consent to participate in the research after all information about the purpose of the research were explained thoroughly [Appendix IV].

### 3.6. Reproducibility of Dental Indices

The study was accomplished to estimate the reproducibility of recording the indices and scores for caries and the plaque and gingival levels. All patients were examined by one examiner (I.N.). The examiner was trained and calibrated to perform precise diagnosis of dental caries scores and plaque and gingival indices scores. The examiner was calibrated by a University Assistant Professor of Paediatric Dentistry (F.Z.) by examining 8 patients aged between 5 to 12 years who attended the Jordan University of Science and Technology Dental Clinics before the commencement of the study, using the World Health Organization (WHO) criteria to assess inter-examiner agreement [WHO, 1997]. Using Kappa test coefficient there was a 98.6 percentage of agreement between the two examiners.
Gingivitis and bacterial dental plaque measurements were registered for eight children by both examiners. There was a 97.7% percentage of agreement between the two examiners. The same patients were examined again after 1 week by the main examiner to assure intra-examiner agreement [Cleaton & Jones, 1989]. There was a total of 98.8 percentage of agreement between the two examinations.

3.7. Oral Examination

Each child participated in this study had a full oral examination; indices for dental caries, gingival plaque and gingivitis in both dentitions were calculated by one dentist using intraoral light and mirror, all disinfection procedures were taken during examination.

For the gingival health status, the gingiva was visually examined for inflammation around every single tooth using a simplified gingival score based on the number of teeth associated with gingival inflammation to give the gingivitis score with either inflamed gingiva around the labial and buccal gingival margins of the tooth investigated (to be given a score=1) or healthy gingiva around the labial and buccal gingival margins of the tooth investigated (to be a given a score =0). The tooth was given a score (1) if parts or all of the buccal cervical gingiva from the mesial papilla to distal papilla appeared reddish, swollen, or if self bleeding from the gingival sulcus was evident at the examination time. Several investigators have used variations of "present or absent" indices which do not consider the severity of gingival inflammation. The observation of whether or not inflammation is present in the gingiva might be a useful approach in clinical studies. Such an index would be simple, reproducible with no invasive methods that could cause unnecessary risk of such a group of patients [Hazen, 1974].

For dental plaque scoring, every single tooth present in the oral cavity was visually examined for bacterial dental plaque deposits to give the plaque score with either present
(to be given a score=1) or not present (to be a given a score =0), using a modification of the O'Leary index [Franco et al., 1996], this simplified scoring system was used previously in previous similar published research by Sonbol et al., 2001.

The Decayed-Missing-Filled Index (DMFT-dmft/ DMFS-dmfs) was used, using the WHO criteria (1987). The caries criteria used were those of the World Health Organization (WHO), which defines caries when “a lesion in a pit or fissure, or on a smooth tooth surface, has an unmistakable cavity, undermined enamel, or a detectably softened floor or wall”. A crown was considered filled, with decay, when it had one or more permanent restorations and one or more areas that were seen to be decayed, A crown was considered filled, without decay, when one or more permanent restorations were present and there was no caries anywhere on the crown. A tooth that has been crowned because of previous decay was recorded in this category. A tooth was considered missing for permanent or primary teeth that have been extracted because of caries. For missing primary teeth, this score should be used only if the subject is at an age when normal exfoliation would not be a sufficient explanation for absence. [WHO, 1997] [Appendix V].

3.8. Statistical Analysis

Statistical analysis was performed using the Statistical Package for Social Science (SPSS) computer software (SPSS 17.0, SPSS Inc., and Chicago, USA).

Descriptive statistics (frequencies, means and standard deviation) were calculated for all the recorded variables for each group. Differences between cases and control groups, for the measured variables and scores were assessed using independent t-test. Significance was pre-determined at 0.05 levels at a confidence interval of 95%.
Chapter Four: Results

4.1. Study Sample

From the names list obtained from The Jordanian Association of Hemophilia and Thalassemia, sixty three names were within the determined age group of this study. Twenty seven children with hemophilia were registered in Rahma Hospital, and thirty six children with hemophilia were registered in three hospitals in Amman. We were unable to contact eleven children of them because of wrong telephone numbers registered in the list, eight patients were severely debilitated and their parents couldn’t reach the examination locations, two patients who were registered in the list were passed away according to their parents when they were contacted, and four patients moved their residence out of Jordan with their families, while the parents of twelve children refused to participate in the study or refused to sign the consent.

The final sample number that could be recruited was twenty six male children diagnosed with hemophilia who were recruited from hemophilia outpatient clinics in Rahma Hospital – Irbid, Al-Basheer Hospital – Amman, Jordan University Hospital – Amman, and Royal Medical Services Clinics – Amman.

A matching number of children who did not have hemophilia were recruited from the Postgraduate and Undergraduate Pediatric and Orthodontic clinics at Jordan University for Science and technology or from the Orthodontic New Patient Assessment Clinic in Jordan University Hospital. Eighteen healthy patients lived in Irbid, six from Amman, one from Zarqa and one from Jerash.
The mean age for the participants in the study group was (9.95 ± 2.18 yrs) while the mean age for those in the control group was (10.05 ± 2.16 yrs). The overall sample age was (10.00 ± 2.15 yrs). The ages ranged from 3 years old to 15 years old in both groups.

Table (4.1) shows the distribution of the two groups among hospitals, residence and age.

**Table 4.1: Distribution of the study group.**

<table>
<thead>
<tr>
<th>Criteria</th>
<th>Study Group</th>
<th>Control Group</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>N(%)</td>
<td>N(%)</td>
</tr>
<tr>
<td><strong>Hospital</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Al-Basheer</td>
<td>4 (16)</td>
<td>__</td>
</tr>
<tr>
<td>Rahma</td>
<td>14 (54)</td>
<td>__</td>
</tr>
<tr>
<td>Royal Services Clinics</td>
<td>4 (16)</td>
<td>__</td>
</tr>
<tr>
<td>JU</td>
<td>4 (16)</td>
<td>__</td>
</tr>
<tr>
<td><strong>Residence</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Amman</td>
<td>9 (34.6)</td>
<td>6 (23.2)</td>
</tr>
<tr>
<td>Irbid</td>
<td>11(42.3)</td>
<td>18 (69.2)</td>
</tr>
<tr>
<td>Jerash</td>
<td>0</td>
<td>1 (3.8)</td>
</tr>
<tr>
<td>Ramtha</td>
<td>1 (3.8)</td>
<td>0</td>
</tr>
<tr>
<td>Zarqa</td>
<td>3 (11.5)</td>
<td>1 (3.8)</td>
</tr>
<tr>
<td>Mafraq</td>
<td>2 (7.8)</td>
<td>0</td>
</tr>
<tr>
<td><strong>Age</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>(Mean ± SD) yrs</td>
<td>(9.95 ± 2.18)</td>
<td>(10.05 ± 2.16)</td>
</tr>
<tr>
<td>Total</td>
<td>26(100)</td>
<td>26(100)</td>
</tr>
</tbody>
</table>

### 4.2 Type and Severity of Hemophilia

Twenty three patients from the study group had Hemophilia type A with a percentage of (88.5%), the other 3 patients had Hemophilia type B with a percentage of (11.5%).

29
Most of the subjects had severe hemophilia (n=18, 69.2%), a lesser number had moderate hemophilia (n=7, 26.9%), and only a case had mild hemophilia (n=1, 3.8%). The frequencies and percentages are shown in the Table (4.2).

**Table 4.2:** The frequencies and percentages for type and severity of hemophilia among the study group.

<table>
<thead>
<tr>
<th>Criteria</th>
<th>N(%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Type of hemophilia</td>
<td></td>
</tr>
<tr>
<td>A</td>
<td>23 (88.5)</td>
</tr>
<tr>
<td>B</td>
<td>3 (11.5)</td>
</tr>
<tr>
<td>Severity</td>
<td></td>
</tr>
<tr>
<td>Mild</td>
<td>1 (3.9)</td>
</tr>
<tr>
<td>Moderate</td>
<td>7 (26.9)</td>
</tr>
<tr>
<td>Severe</td>
<td>18 (69.2)</td>
</tr>
<tr>
<td>Total</td>
<td>26 (100)</td>
</tr>
</tbody>
</table>

**4.3. Intraoral Examination**

**4.3.1. Periodontal Assessment**

4.3.1.1. Bacterial Plaque

When the two groups plaque scores were compared, the statistical analysis revealed that the subjects in the study group had higher plaque scores (3.62 ± 4.54) in comparison to the control group (3.46 ± 3.22) in primary teeth, however, this was statistically insignificant as the probability was >0.05 (p= 0.888 for primary teeth). In contrary, plaque scores in permanent teeth for the study group (6.15 ± 6.25) was lower than that of the control group (6.85 ± 5.81), but this difference was statistically in-significant (p=0.681 for secondary teeth). Table (4.3) shows the bacterial plaque scores’ comparisons between the two groups.
Table 4.3: Bacterial Plaque Scores: Study and Control groups.

<table>
<thead>
<tr>
<th>Score</th>
<th>Study group</th>
<th>Control group</th>
<th>P value</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Mean ± Sd</td>
<td>Mean ± sd</td>
<td></td>
</tr>
<tr>
<td>Plaque score</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Primary Teeth</td>
<td>3.62 ± 4.54</td>
<td>3.46 ± 3.22</td>
<td>0.888</td>
</tr>
<tr>
<td>Permanent Teeth</td>
<td>6.15 ± 6.25</td>
<td>6.85 ± 5.81</td>
<td>0.681</td>
</tr>
</tbody>
</table>

4.3.1.2. Gingival Health

As for the gingival health, the control group had higher gingival scores in both primary and permanent teeth; for primary teeth the control group gingival score was \( (3.92±3.11) \) compared to \( (3.50 ±5.12) \) in the study group, while for permanent teeth it was \( (8.38±7.28) \) compared to lower readings in the study group \( (7.54±7.12) \). In both comparisons there was no significant difference as the p value was > 0.05 \( (p=0.720) \) for primary teeth and \( (p=0.674) \) for secondary teeth. The comparison between the two groups in the gingival index scores are shown in Table (4.4).

Table 4.4: Gingival Index Scores: Study and Control groups.

<table>
<thead>
<tr>
<th>Index</th>
<th>Study group</th>
<th>Control group</th>
<th>P value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Gingival Index Score</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Primary Teeth</td>
<td>3.50 ± 5.12</td>
<td>3.92 ± 3.11</td>
<td>0.72</td>
</tr>
<tr>
<td>Permanent Teeth</td>
<td>7.54 ± 7.12</td>
<td>8.38 ± 7.28</td>
<td>0.674</td>
</tr>
</tbody>
</table>

4.3.2. Dental Caries Indices

There was no significant difference between cases in the study and control groups in the DMFS scores; as the p value was 0.681; the median of DMFS score in the study group= 3.08±3.46 and in the control group it was 2.69±3.21. Upon further analysis, it was shown that all individuals in the control group had MS=0, while for the control group the
median value of MS=0.27 ± 1.37, however, this was statistically in-significant. The study group had a DS score =2.85 ± 3.09 which was higher than Ds score in the control group DS = 1.92± 2.74, but also this was statistically in-significant. There was a larger difference between the median values of FS score; in the study group FS= 0.23± 0.71, while in the control group FS= 1.04±2.22, but it was in-significant since the p value equaled 0.083 (Table (4.5)).

Table 4.5: Decayed, Missing, and Filled Surfaces Score in Permanent Teeth.

<table>
<thead>
<tr>
<th>Index</th>
<th>Study group</th>
<th>Control group</th>
<th>P value</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Mean ± SD</td>
<td>Mean ± SD</td>
<td></td>
</tr>
<tr>
<td>Caries index</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>DS</td>
<td>2.85 ± 3.09</td>
<td>1.92 ± 2.74</td>
<td>0.260</td>
</tr>
<tr>
<td>MS</td>
<td>0</td>
<td>0.27 ± 1.37</td>
<td>0.322</td>
</tr>
<tr>
<td>FS</td>
<td>0.23 ± 0.71</td>
<td>1.04 ± 2.22</td>
<td>0.083</td>
</tr>
<tr>
<td>DMFS</td>
<td>3.08 ± 3.46</td>
<td>2.69 ± 3.21</td>
<td>0.680</td>
</tr>
</tbody>
</table>

There was no significant difference neither in the DMFT scores between the study and control group, nor in its sub-scores (DT, MT, FT). The DMFT score in the study = 2.08 ± 2.10, and for the control group it was 1.85 ± 2.27. (Table (4.6)).

Table 4.6: Decayed, Missing, and Filled Teeth Score in Permanent Teeth.

<table>
<thead>
<tr>
<th>Index</th>
<th>Study group</th>
<th>Control group</th>
<th>P value</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Mean ± SD</td>
<td>Mean ± SD</td>
<td></td>
</tr>
<tr>
<td>Caries index</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>DT</td>
<td>1.88 ± 1.97</td>
<td>1.31 ± 2.09</td>
<td>0.311</td>
</tr>
<tr>
<td>MT</td>
<td>0</td>
<td>0</td>
<td>-</td>
</tr>
<tr>
<td>FT</td>
<td>0.19 ± 0.49</td>
<td>0.54 ± 0.90</td>
<td>0.093</td>
</tr>
<tr>
<td>DMFT</td>
<td>2.08 ± 2.10</td>
<td>1.85 ± 2.27</td>
<td>0.705</td>
</tr>
</tbody>
</table>
On the other hand there was a significant difference in the dmfs scores between the two groups; in the study group it was \(8.23 \pm 7.78\), it was obviously lower than the median score in the control group which was \(15.08 \pm 11.87\) \((p=0.017<0.05)\). A highly significant difference was shown upon further analysis in fs score since \(p\) value= 0.001. There was insignificant difference in the ds and ms scores (Table (4.7)).

<table>
<thead>
<tr>
<th>Table 4.7: Decayed, Missing, and Filled Surfaces Score in Primary Teeth.</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Index</strong></td>
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<tr>
<td></td>
</tr>
<tr>
<td>Caries index</td>
</tr>
<tr>
<td>ds</td>
</tr>
<tr>
<td>ms</td>
</tr>
<tr>
<td>fs</td>
</tr>
<tr>
<td>dmfs</td>
</tr>
</tbody>
</table>

No significant difference was present in dmft score; median value of dmft score was \(3.85\pm3.68\) in the study group, while in the control group it was \(4.81\pm2.95\), \(p\) value=0.304. But further analysis has shown significant difference in ft score; in the study group it was \(0.54\pm0.86\), which was much lower than that of control group \((ft=1.50\pm1.70)\), \(p\) value = 0.013. No significant difference was shown in the dt and mt scores between the two groups (Table (4.8)).

<table>
<thead>
<tr>
<th>Table 4.8: Decayed, Missing, and Filled Teeth Score in Primary Teeth.</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Index</strong></td>
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<tr>
<td></td>
</tr>
<tr>
<td>Caries index</td>
</tr>
<tr>
<td>dt</td>
</tr>
<tr>
<td>mt</td>
</tr>
<tr>
<td>ft</td>
</tr>
<tr>
<td>dmft</td>
</tr>
</tbody>
</table>

All patients examination details are shown in Appendix VI.
Chapter Five: Discussion

By reviewing the literature, minimal information about the oral health of this group of patients is noticed [Zeibolz et al., 2011].

5.1. Study Design

Twenty six children with severe, moderate, and mild hemophilia were recruited from reference hospitals for hemophilia in Jordan. The entire sample was males; and this was expected since hemophilia disease is an X-linked recessive trait disorder which occurs mostly in males or in very rare cases in homozygous females [Aster & Robbins, 2003]. Two females within the age group of this research were registered in the lists of hemophilic patients but unfortunately they were unable to participate in the study because of long distance traveling in order to reach the examination clinics in JU hospital clinics or JUST postgraduate clinics as their families claimed. The study group children were matched in age and gender with the control group; who were mainly selected from the "Orthodontic and Trauma postgraduate and undergraduate clinics" patients in Jordan University for Science and Technology, or from "New assessment orthodontic clinic" in Jordan University hospital. Those options removed bias that could be resulted if children who were asked to visit the dentist for treatment of caries were examined.

Hemophilia A, von Willebrand disease and Glanzmann thrombasthenia are the most frequently seen inherited bleeding disorders in Jordan [Awidi, 2005]. The data for bleeding disorders stem from cases presented to hospitals or laboratories. Therefore, data concerning patients with hemophilia’ prevalence are scarce [Awidi et al., 2010].
According to the Jordanian National Registration Record in 2012 only 300 cases were registered as patients with hemophilia, on the other hand more than 200 cases are expected to be present but unregistered. This explains the small sample of this study when the inclusion criteria with the defined age group were determined.

A simplified index was used to assess the gingival score and plaque which depends on visual assessment only under dental unit light since the subjects' bleeding disorder could be affected in case of using a sharp dental probe and may cause unnecessary bleeding from the gingiva. A similar study has used simplified indices to avoid the risk of bleeding in such patients [Sonbol et al., 2001].

5.2. Gingival Health

Gingivitis is a result to the gathering of oral bacteria in the gingival sulci; usually diagnosed by clinical signs and symptoms [Summers, 2009]. Gingivitis is expressed by redness, swelling and bleeding on gentle probing of the gingival sulcus [Summers, 2009].

In previous publications in 13–14-year-old Jordanian children from the north it was reported that Plaque Index scores were 1.82 and 1.63 during 1993 and 1999 respectively. Furthermore, gingival scores reported in these school children in 1993 and 1999 were 1.89 and 1.67 respectively [El-Qadri et al., 2006].

In our study there was no significant difference in the gingival health status between children with hemophilia and healthy children, neither in primary nor in permanent dentition; both of them had relatively high plaque and gingival scores. According to some workers those scores are a reflection to the dental education level of parents which is comparably low in developing countries [Davies et al., 2000].

A similar study conducted in Northern Ireland also found no difference between the children affected with hemophilia and healthy matched control group but unlike the
present study the plaque and gingival scores were low; this was attributed to the fact that
dental care of this category of patients is carried out at the regional treatment centre and
forms an integral part of their medical care [Boyd et al., 1997]. Similar results were
reported in a study on British children, but more interestingly healthy children had
significantly higher plaque scores in permanent dentition [Sonbol et al., 2001]. On the
other hand, Alpkilic et al., 2009 reported in a similar study design higher gingival index
scores for children with hemophilia in relation to healthy children.

Poor oral health education in developed countries can explain the reason for high
plaque and gingival scores in the study group. Some workers in an Egyptian study have
reported marked improvement in oral hygiene after reviewing patients six months after the
first examination date and giving them proper instructions regarding dental and gingival
care [Kabil et al., 2007]

It is important to emphasize that the mean for gingival scores for the whole sample
was higher than that reported in 1993 and 1999 by Taani et al. in North Jordan, which
highlights the importance of oral hygiene instruction programs and dental health education
for school children in Jordan in general, and for the medically compromised children
especially those with inherited bleeding disorders.

5.3. Dental Caries

Dental caries is the most common disease of children and considered a significant
oral health problem worldwide for all age groups [Young et al., 2010].

Under normal conditions, oral fluids are supersaturated with respect to
hydroxyapatite. However, when the pH of oral fluid reduces below the critical pH, the
supersaturation with respect to hydroxyapatite is replaced by undersaturation, resulting in
the mineral loss (demineralization) of enamel. When the pH reduction is due to bacteria-
derived acid production, this mineral loss is called caries [Watanabe et al., 2012].
Because of the thin enamel in deciduous dentition the caries spreads faster than teeth of adults, which can result in toothache, teeth destruction, and may be systemic infection [Christian et al., 2011]. Newly erupted permanent teeth are also more prone to get demineralized faster due to the fact that the crystal structure of hydroxyapatite in enamel is immature when a tooth erupts, and it gradually matures after tooth eruption because of minerals in oral fluid, water, food, etc [Watanabe et al., 2012].

Caries risk indicators are variables that have direct influence to the process like microflora, or indirect effect like socioeconomic status and other things and include those variables that may be considered protective factors [AAPD].

Some authors have investigated the prevalence of dental caries in Jordan; 16–8% of them had missing or filled teeth with a DMFT of 3.13 according to El-Qadri et al., 2006, previous studies have revealed that the DMFT scores in 13–15-year-old school children was between 4 and 5 [Taani et al., 1997].

No significant difference was observed in the DMFS or DMFT scores in our present study. This may be attributed to the fact that most children were having newly erupted permanent teeth with low scores (DMFS=2.88) and (DMFT= 1.96). A study on British children have reported dental caries measures were greater in the control children in comparison with diseased group, however, all caries scores were less than the present study in both groups (DMFS=0.8) and (DMFT= 0.7) [Sonbol et al., 2000].

The DMFT was almost similar to some patients with chronic diseases. The DMFT for children with clefts in Jordan was 1.68 [Hazza et al., 2011], children with unilateral cleft lip and palate was 1.9 [Lucas et al., 2000], and 0.6 for children with phenylketonuria [Lucas et al, 2000]. In opposite, the DMFT was much less than Polish children, 5.8 [Blaszczechak, 1999], lower than that reported in Indian hemophilic children.
2.34 [Saxena and shashikiran, 2010], and lower than Egyptian hemophilic children 2.95 [Kabil et al., 2007].

On the other hand, a significant difference was evident between the two examination groups in the results of dmfs scores; the control group had higher dmfs scores, but further analysis has revealed that the higher dmfs scores were attributed to the higher (f) scores. This raises an important issue; that control children had more accessible dental treatment which explains higher (fs) scores. The same conclusion was confirmed when the control group had higher dmft scores with significantly higher (ft) scores. Unlike the results reported in other studies the main component of the dental caries score in primary teeth was (dt) which refers to decayed primary teeth [Kabil et al., 2007].

Oral care for hemophiliacs does not have a strategic significance in developing countries, as they usually get minimal oral health treatments, although these dental untreated dental problems can lead to major problems regarding their medical condition [Davies et al., 2000].

5.4. Conclusion

Dental care education is mandatory for children in order to keep good oral health. When this is not applied there is a greater chance for having oral health complications during life. However, dental care in most developing countries including Jordan is minimal, and this was obvious through the relatively high plaque scores, gingivitis scores, and dental indices score in our study. This is most probably due to the high cost of dental services; in addition, oral health education has less importance than other developed countries. However, those indices were lower than similar group of children in comparison with other countries or compared with other medically compromised children, and this might be due to the role of education programs—although minimal—directed to this
specific group of patients through their medical centers. Some researchers thought that higher gingivitis and caries indices results in children with hemophilia were a consequence of neglected or insufficient tooth brushing habit as most of them are afraid from gingival bleeding as a consequence of brushing [Alpkilic et al., 2009]. In general, the noticed differences might be due to economic status and differences in provision of dental care [Zeibolz et al., 2011].

We concluded in this study that children with hemophilia have difficult access to dental treatment related to many barriers that need to be clarified through other study designs.

5.5. Recommendations

- A prospective study with formulated diet sheets and recording of practices and oral hygiene habits is recommended to specifically control the etiological factors of dental caries and gingivitis in children with hemophilia.
- Also more studies are required to investigate the multifactorial etiology of dental caries in hemophilic children like socioeconomic factors; dietary habits, use of fluorides, plaque control, saliva and bacteria.
- Dental care for all medically compromised children and especially for children with hemophilia should form an integral part of their medical care. Medical practitioners and Hematologist should participate in oral health education programs and communicate well with dental practitioners to facilitate case referral and proper dental treatment.
5.6. Limitations of the Present Study

- The sample size of this study is considered small but since Hemophilia is considered a rare inherited bleeding disorder, and since the whole sample of patients who were registered as patients with hemophilia according to the Jordanian National Registration Record in 2012 was only 300 cases, the sample of this study could be considered representative to the whole Jordanian sample when defined age group was determined. In addition, many other similar studies conducted in other countries had almost similar sample size [Sonbol et al., 2001; Alpkilic et al., 2009] or even less than our sample [Zeibolz et al., 2011].

- Radiographs were not present to complete clinical diagnosis of dental caries, and this was attributed to many reasons; the main reason was that most of the parents of the study sample refused to do the examination anywhere except in their hemophilia outpatient clinics related to Rahma Hospital where there was no dental panoramic or x-ray machines, fewer number of parents refused to sign the consent and participate in the study if x-rays were to be taken to their children. And since the sample size was already small for this group of patients, it was impossible to exclude all these cases from the study sample, so this might give underestimation of the actual caries level in the study sample.
References:


National Hemophilia Foundation web site www.hemophilia.org / NHFWeb / MainPgs / MainNHF Accessed on June 8th 2013.


APPENDICES

APPENDIX I

Ethical approval from the IRB at JUST
Evaluation of the oral and dental health of children with hemophilia in Jordan

يرى إعلامكم بموافقة لجنة البحث على الإنسان على إجراء البحث العلمي المشار إليه أعلاه، على أن يتم التقيد بالشروط التالية:

1. الحفاظ على سرية المعلومات وأن لا تستخدم إلا لغايات البحث العلمي.
2. وضع نسخة من نموذج التقويم الخاص من المرضى المشاركون في ملف الطبيب، والاحتفاظ بنسخة أخرى مع الباحث لأغراضها عند الحاجة، علمًا وأنه سيتم التأكد من ذلك من خلال التدقيق على ملفات المرضى المشاركون في الدراسة.
3. تزويدها بأسماء المرضى المشاركون في البحث (أرقام الملفات الطبية لهم) وتوقيع تكساليف الممارسات عن المشاركون.
4. تزويدها بنسخة من نتائج البحث.

وتفضلوا بقبول فائق الاحترام،

رئيس لجنة البحث على الإنسان

الاستاذ الدكتور محمود الشيبان


APPENDIX II

University of Jordan permission
الأستاذ الدكتور مدير مستشفى الجامعة الأردنية

تحية طيبة وبعد،

أرجو التكرم بالموافقة والإعصار للمعينين لتسهيل مهمة الطالبة إيمان سعيد محمد
تزال ورقها الجامعي (2013-6) تخصص طب أسنان الأطفال لإجراء
فحص المرضى لأغراض البحث العلمي المسمى:

"تقييم حالة صحة الفم والأسنان عند الأطفال من مرضى هيموفيليا الدم في
الأردن"

حيث أن الدراسة تتضمن ملء استبيان فحص الفم وعمل صور أشعة.

شكراً ونと一緒に لكم مساهمتنا.

رحمه

رئيس الجامعة

الذين يجتمعون

ونفضل عطوفتك بقبول فائق الاحترام.

P.O. Box (3030) Irbid 22110 Jordan. Tel (962-2) 7201000 - Fax (962-2) 7095123
E-mail: prsdy@just.edu.jo
website: http://www.just.edu.jo

APPENDIX III

Ministry of health hospitals permissions
APPENDIX IV

Consent Form
Consent Form

Mo'awafa ʿala l’ashtarak ʿalai baytibbi

Consent Form to Participate in Research

Ana al-mawqoofi adanah .....

Qiviim halata safah al-fam w al-a{sanas anf al-adfalli min mursi himofiliba adllat al-ardan

al-talabba

aim siyid nizal tuht Enhara daktorat fada sahaya w Enhara daktorat hawazn seil.

al-bagag sof ipatim fiqasureri alfam w sof asura alasanas w alwoghe un aldoom w hasab ma yirehna tibeb

al-muwajha.

Lakd ta shraqibiyat al-bagag w adhafaha w tafhemet al-mawzour ka'mala un ana la ahimalia tibiyat maliai jarra dakh wakd ta

Elaamni bani maraarkti fi zahar bagag xibaria w talawiyah wana he la yikhdaxi ya irjaarida la qarrrt ana a sharak ou

Ea sharakti fi al-bagag tha qarrrt ana aqof ma'sahmat.

Qawafqa un a sharak telafi jasenn hawzah dersa wala irjaarSidat asura alasanas yadnuriy.Wa

asam alfali:

Towiq wa-li almar:

al-taqar:
APPENDIX V

Examination sheets used for gingival, plaque, and caries scores

**Plaque scores-**
Two gingivally related points of each tooth (mesiobuccal, distobuccal) were visually examined for bacterial dental plaque deposits to give the plaque score, using a modification of the O'Leary index. (0= no plaque deposit) (1= plaque deposits present).

<table>
<thead>
<tr>
<th>E</th>
<th>D</th>
<th>C</th>
<th>B</th>
<th>A</th>
<th>A</th>
<th>B</th>
<th>C</th>
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</table>

**Gingival index scores –**
Simplified gingival index based on the number of teeth associated with gingival inflammation. (0= no inflammation) (1 = gingival inflammation)

<table>
<thead>
<tr>
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### Caries index

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- **dmfs:** ...............  
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- **DMFT:** .....................  
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- **MS:**.....  
- **FS:**.....  
- **DT:**.....  
- **MT:**.....  
- **FT:**.....  

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52
## APPENDIX VI

### Hemophiliac Patients Data Histogram

<table>
<thead>
<tr>
<th>Gingival score in secondary</th>
<th>Gingival score in primary</th>
<th>plaque in secondary teeth</th>
<th>plaque in primary</th>
<th>no of permanent teeth present</th>
<th>no of primary teeth present</th>
<th>dmft</th>
<th>ft</th>
<th>mt</th>
<th>dt</th>
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52
تقييم حالة صحة الفم و الأسنان عند الأطفال من مرضى هيوموفيليا الدم في الأردن

إعداد: إيمان سعيد نزال

الملخص

المقدمة: تعتبر هيوموفيليا واحدة أكثر أمراض تزف الدم شيوعاً في الأطفال، فيما أن صحة الفم والأسنان لا تعتبر إجراء مهمًا للمرضى الذين يعانون من هيوموفيليا خاصة في الدول النامية، إلا أن هذا المر قد يسبب لديهم المضاعفات التي يمكن أن تتفاعل مع صحتهم.

الأهداف: إن الهدف من هذه الدراسة هو قياس مدى انتشار تسوس الأسنان، والتهابات اللثة والبلاك عند الأطفال المصابين بالهيوموفيليا وتطبيق ذلك إلى الأطفال الأصحاء في عينة من الأطفال الأردنيين.

أسلوب ومواد: تم دراسة هذه الحالات من مناطق الشمال والمناطق الوسطى في الأردن، تم معاناة وفحص ستة وعشرين من الأطفال الذين يعانون من الهيموفيليا من عدة مستشفيات في هذه المناطق في الأردن، وقد تم متابعتهم مع الأسنان المصنوعة من حيث الجنس والعمر. سجلت مؤشرات تسوس السنان حسب DMFS/DMFT للأطفال الذين يعانون من هيوموفيليا، مما أدى إلى قياس درجات البلاك والتهاب اللثة.

النتائج: سجل الأطفال الذين يعانون من الهيموفيليا درجات أقل من حيث انتشار البلات من الأطفال الأصحاء في الأسنان الدائمة بينما كانت درجاتهم أعلى قليلاً في الأسنان اللبنية، أما النهايات اللثة أيضاً فقد كانت أقل عند الأطفال الذين يعانون من الهيموفيليا في الأسنان اللبنية والدائمة. أما بالنسبة للتسوس في السنان الدائمة فقد كانت المؤشرات أعلى في الأطفال الذين يعانون من الهيموفيليا في الأسنان الدائمة ولكن الفرق لم يكن حاسمًا إحصائيًا، أما بالنسبة للأسنان اللبنية فقد كانت مؤشرات النسوس أعلى عند الأطفال الذين يعانون من هيوموفيليا، إلا أن مزيد من التحليل أوضح أن الفرق كان بسبب درجات أعلى بكثير من الأسطح المملوءة بالحشوات والتي قد عانوها عند هؤلاء الأطفال.

الخلاصة: العلاج بأمان للأطفال الذين يعانون من الهيموفيليا في معظم البلدان النامية بما فيها الأردن، هو بالحد الأدنى يمكن برامج الوقاية للأسنان وتقييمها بشكل دقيق يقلل كثيراً من الحاجة للعلاج وتجنب الحاجة لقطع الأسنان عند هذه الفئة من الأطفال تجديداً.