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# Haemophilia: Pediatric Dentistry Perspective: A Review

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## Abstract

Hemophilia is an X-linked congenital bleeding disorder caused by a deficiency of coagulation factor VIII (FVIII) (in hemophilia A) or factor IX (FIX) (in hemophilia B). The deficiency is the result of mutations of the respective clotting factor genes. It affects males on the maternal side. The incidence is estimated to be 1 in 5,000 males for hemophilia A and 1 in 30,000 males for hemophilia B. The clinical manifestations are easy bruising, joint and muscle hemorrhage. Children with haemophilia rubber cup prophylaxis and supragingival scaling may be safely performed without replacement therapy. Factor replacement before frenectomy and other periodontal surgeries. Electrosurgery is done because of the possibility of continued bleeding. Most restorative procedures on primary teeth can be successfully completed. Thin rubber dam is preferred. Wedges and matrices can be used conventionally. A pulpotomy or pulpectomy is preferable to extraction but instrumentation in periapical area should be avoided. Nonvital teeth should be obturated 2 to 3 mm short of apex. Use retraction cords during crown preparation and antibiotic prophylaxis before extraction. Infiltration anesthesia can generally be administered without replacement therapy. A minimum of a 40% factor correction is mandatory before block anesthesia. In the absence of factor replacement, periodontal ligament (PDL) injections may be used. Use of topical hemostatic agents such as bovine thrombin, microfibrillar collagen hemostat gel foam, absorbable oxidized cellulose etc. Arch wires should be secured with elastic bands. Removable appliances are used with Adams or similar clasps without free ends and preformed bands and brackets are preferred. When general anesthesia is considered, oral intubation is preferred over nasal intubation, Intramuscular injections should be avoided. This article discuss about etiology, clinical features and management of children with hemophilia.

***Keywords:*** *Bleeding Disorder; Children; Dental Management; Hemophilia; Oral Health*

## Abbreviations

F VIII: Factor VIII; F IX: Factor IX; PDL: Periodontal Ligament; F VIII: Factor VIII; CNS: Central Nervous System; AHG: Antihemophilic Globulin; PTC: Plasma Thromboplastin Component; PTA: Plasma Thromboplastin Antecedent; AHF: Anti Hemophiliac Factor; EACA: Epsilon Amino Caproic Acid; NSAIDs: Non-Steroidal Anti-Inflammatory Drugs

## Introduction

Hemophilia is an X-linked recessive disorder caused by a deficiency in blood coagulation factors. It is the second most common coagulation disorder after von Willebrand disease. Von Willebrand disease is prevalent in approximately 1% of the general population and affects both males and females with equal frequency [1]. Hemophilia A is characterized by a deficiency of factor VIII (F VIII), whereas hemophilia B is caused by a deficiency of factor IX (F IX). The incidence is estimated to be 1 in 5000 males for hemophilia A and 1 in 30,000 males for hemophilia B [2]. Since hemophilia is X-linked, men typically express the disease, whereas women tend to be asymptomatic carriers. However, both F8 and F9 genes are prone to new mutations, and as many as 1/3 of all cases are the result of spontaneous mutation where there is no prior family history [2].

The clinical characteristics of both types of haemophilia are similar: spontaneous or traumatic haemorrhages; muscle haematomas; haemophilic arthropathy caused by recurrent bleeding into target joints; and bleeding into the central nervous system (CNS). Hemarthroses are first seen when the child learns to walk. If the joint hemorrhages go untreated, it can lead to permanent disability.

Following the primary stages of control of bleeding from a wound by vasoconstriction and formation of a platelet plug, the blood clotting mechanism initiates. This complicated reaction involves many factors (Table 1), and deficiency in one of them can break the chain.

| | |
|---|---|
| Factor I | Fibrinogen |
| Factor II | Prothrombin |
| Factor III | Thromboplastin |
| Factor IV | Calcium |
| Factor V | Pro accelerin, accelerator globulin, or labile factor |
| Factor VI | Activated labile factor (sometimes omitted) |
| Factor VII | Stable factor or pro convertin |
| Factor VIII | Antihemophilic factor, antihemophilic globulin (AHG) |
| Factor IX | Plasma thromboplastin component (PTC) |
| Factor X | Stuart-power factor |
| Factor XI | Plasma thromboplastin antecedent (PTA) |
| Factor XII | Hageman factor |

***Table 1:*** *Factors concerned in blood coagulation.*

Hemophilia is inheritable, the commonest of them being classic hemophilia or deficiency of the antihaemophilic factor and accounts for three-quarters of cases. Christmas disease was differentiated from true hemophilia relatively recently when laboratory techniques became sufficiently developed. Hemophiliacs generally have a prolonged coagulation time and show an abnormal tendency to bleeding from wounds, tissues, and joints [3].

### Hemophilia A

(Classic hemophilia, deficiency of anti haemophilic factor (AHF), anti haemophilic globulin (AHG) or a factor VIII).

True hemophilia is a deficiency of factor VIII, mainly affecting males, and inherited as a sex-linked recessive character. The defect is carried on the X chromosome so that a hemophiliac father and a normal mother will have normal sons and carrier daughters, while a normal father and carrier mother will have normal and affected sons and normal and carrier daughters.

The possibility of the combination of a hemophilic father and a carrier mother is very slight, so that the chances of a female with both X chromosomes being defective, and therefore clinically affected, appear to be remote but has occurred. A female carrier may have some degree of deficiency of factor VIII and rather prolonged coagulation time, though rarely a bleeding tendency of real clinical importance. More than a quarter of the cases appear to have no family history of the defect, and through some of these must be due to smallness of numbers in the family; nevertheless, the rate of mutation producing primary cases seems relatively high.

The degree of severity affecting members of the same family tends to be very similar, and some families will have severe cases while other families have mild ones. Clinical problems arise in patients with 30% or less factor 8, but rarely with levels above this. However, the clotting time of whole blood may be familiar with only 5% or less, and the patient's history and clinical condition must be considered and make a possible diagnosis. Those with mild hemophilia have a 30% and lower and may remain undiscovered until adulthood though they may give a history of a dental extraction in childhood in which the bleeding was prolonged but not so severe as to warrant hospital help. The more severely afflicted who have little or no factor 8, suffer repeated hemorrhagic episodes from early childhood, particularly when learning to walk when falls are frequent and extensive bruising, or a hematoma may result from a trivial blow. External bleeding from an impact on a nose or front teeth is a regular occurrence in a toddler.

Small cuts and scratches are usually of no importance as the bleeding usually stops, but more significant wounds require some control. Topical measures are used first and include the application of pressure, cold, fresh or powdered thrombin, epinephrine, and immobilization of the affected part. Suturing should be done only in the hospital, and never as a first-aid measure superficial bruises are usually of no clinical importance, but deep hematomas may be severe. In muscle, they can cause significant pain and swelling and lead to deformity, and in the abdomen and neck, they become hospital emergencies.

Apart from the main difficulty of bleeding, the problems are of hemorrhage into joints. Occurs sooner or later in all the more severe cases, repeated episodes lead to alkalosis and deformity. When it happens, the patient is in extreme pain, and there are swelling and limitation of movement. Another possible effect of internal hemorrhage is the development of blood cysts, causing bone erosion.

Treatment in the hospital to control hemorrhage may include whole blood, fresh frozen plasma, or AHG fraction. Whole blood is usually given when there has been severe blood loss, and replacement is needed; otherwise, fresh frozen plasma or AHG used [4-6].

| | | |
|---|---|---|
| Severe | < 0.01 IU/mL or <1% | Spontaneous joint, muscle and internal bleeding; excessive bleeding with trauma or surgery |
| Moderate | 0.01-0.05 IU/ml, or 1% to 5% | Bleeding into joints or mimics with minor trauma; excessive bleeding with surgery |
| Mild | 0.05 - 0.35 IU/ml or 5% to 35% | No spontaneous bleeding; delayed onset bleeding after trauma or surgery or dental extractions |

***Table 2:*** *Classification of hemophilia.*

## Christmas disease (Rosenthal syndrome)

Hemophilia B deficiency of factor IX or plasma thromboplastin component (PTC).

Deficiency of factor IX is a defect transmitted as the sex-linked recessive character precisely the same way as classic hemophilia, but the female carriers have a greater tendency to some degree of bleeding abnormality. Clinically the effects are the same as those of factor VIII deficiency and indistinguishable from them in the treatment of hemorrhage. However, a concentrate of factor IX is not ordinarily avail-

able, and plasma appears to be the best source for raising the factor's blood so that those patients present a challenging clinical problem. Patients with mild symptoms may have about a 5 percent level of the blood, while the severely affected have none detectable. The latter tends towards spontaneous bleeding, while the former merely have prolonged bleeding and considerable bruising after trauma [1].

### Factor XI deficiency (Plasma thromboplastin antecedent (PTA) deficiency, hemophilia C)

It is a deficiency in which there is a prolonged coagulation time as in lack of factor VIII and IX, but the mode of inheritance is different. It is transmitted as a dominant character, not sex-linked, and a defective gene from either parent results in the child being affected. The children of a deficient parent can expect to numerically half regular and half affected. The degree of severity in members of the same family is very variable, but the clinical effects are similar to those of deficiency of factors VIII and IX, though they tend to be less severe and vary in degree from time to time in the same patient. The blood level of factor XI can be improved by the administration of plasma to treat hemorrhage.

### Other clotting factors deficiencies

Deficiencies of other factors in the clotting mechanism of the blood occur but are very rare.

Afibrinogenemia or factor I deficiency appears to be a non-sex linked recessive character in which the final phase of the coagulation process is prevented. Prolonged bleeding occurs as in other flaws, but hemorrhage into joints is not a feature. The administration of fibrinogen can bring about hemostasis.

Hypoprothrombinemia or deficiency of prothrombin or factor II occurs in either sex. Still, it is scarce in the congenital form, most of the cases being of the acquires type in which there is liver dysfunction or lack of vitamin K or both. In the adult, it may associate with the use of the coumarin group of anticoagulants. Coagulation time is prolonged, and there may be severe hemorrhagic episodes. Treatment is by the administration of vitamin K where this is needed, and whole blood transfusions [2].

Factor V deficiency is inherited as a recessive character or may be acquired in association with liver dysfunction and the other diseases. As with different types, bleeding is prolonged and spontaneous hemorrhage may occur, but joint complications not expect. Treatment of hemorrhage is by the administration of plasma.

Factor VII deficiency is an inherited recessive character. The factor appears to be formed in the liver and is therefore also reduced in the presence of liver damage, vitamin K deficiency, or the administration of coumarin drugs. Patients have abnormal bleeding and bruising, spontaneous hemorrhage, and some joint problems treatment of hemorrhage in the congenital type by the whole blood or plasma administration.

The deficiency of factor X or Stuart power factor was differentiated from that factor VII only recently by laboratory techniques. It is transmitted as an incompletely recessive character, and while the homozygote is affected severely, the carrier may have the condition to a mild degree. The factor is dependent on vitamin K, and where this is lacking, the deficiency is more severe [1,2,7].

### Von Willebrand's disease (Pseudohaemophilia)

It is a defect of hemostasis, which is, in many respects, distinct from the others of this group. It is inherited as a dominant character equally by both the sexes. There is a prolonged bleeding time, unlike hemophilia, but a standard clotting time and platelet count. There is a defect of the capillary structure so that a damaged vessel continues to ooze for an extended period, and there may be a platelet defect besides.

The condition is characterized by spontaneous bleeding from mucous membrane surfaces and excessive hemorrhage from minor trauma, but hemorrhage from minor trauma but hematomas and bleeding into the joints are not a feature. There is variability in the bleeding

into joints that are not a feature. There is variability in bleeding tendency in any particular patient at different times. Where the deficiency of factor VIII is not significant, local measures to arrest hemorrhage are usually adequate, particularly pressure, provided the lesion is accessible. Otherwise, control is established by raising the level of factor VIII in the blood with plasma or AHG concentrate.

Children who are severely affected with this condition must be regarded as hemophiliacs as they have factor VIII deficiency and what has been said concerning the general care of hemophiliacs also applies to these patients. However, all cases, even the mild ones, must be so regarded soar the dental care because of the hemorrhage potential in the event of surgery and deep injections [8,9].

## General care of children with haemophilia

On the diagnosis, each patient issued an official information card giving the diagnosis, blood group, and the hospital's address of the or center, which looks after the patient. In developed countries, many hemophilia centers set up by the ministry of health and medical research council, which provides a laboratory service for diagnosis, clinical help, and advice for patients and their parents and medical practitioners who have hemophilia patients. Treatment of such patients generally, including surgery, is carried out in many large general hospitals.

The parents are advised to have the usual immunizations carried out since they involve the injection of only a small quantity of fluid and rarely give trouble provided firm finger pressure is applied to the site of injection for five minutes. Injections of larger quantities such as that gamma globulin against measles are potentially dangerous and therefore contraindicated. Any injections should be by the oral route or intravenously, and never intramuscularly. Such patients should never take aspirin or products containing aspirin as it has an irritant effect on the intestinal mucosa and can cause severe gastrointestinal hemorrhage, and it also harms homeostasis. The alternatives to aspirin are panadol, codeine; paracetamol; codeine phosphate [9,10].

A hemophilic child's parent has a difficult problem to face as they must guard him against many dangers which are irrelevant to an average child, and yet not overprotect him. He must not often check that he feels thwarted but has as much freedom as can reasonably be allowed. The maintenance of discipline can be difficult, and occasionally such a child is unmanageable because of overindulgence.

Education may become a problem for some of these children. The mild cases will attend regular school and have little difficulty.

However, the severely afflicted will find such a school too rough and be upset at not playing organized games with friends. Because of his condition, he will be absent for considerable periods and be unable to keep up with his class. Such a child may attend a small local school where he can have individual attention, a school for disabled children near enough to attend daily, or a boarder.

Physical activity is important and can contribute to better coordination, endurance, flexibility and strength [12,13].

There is a hemophilia society to which many parents of affected children belong and share their difficulties and experience. The organization also gives advice and does investigations on social problems relating to the disorder [8,9].

## Oral condition

There are no special distinctive features associated with haemophilia. Their oral hygiene often is low since they may not brush their teeth regularly for fear of initiating bleeding, and of course, this neglect tends to lead to gingivitis and even more painless bleeding of the gums.

The dental caries rate may be relatively high in some patients. Their children are often consoled with sweets for being prevented from doing something undesirable. Meals may be mainly composed of carbohydrates because of the anxiety of causing Gingival bleeding by hard or fibrous foods. These factors and the lack of oral hygiene all predispose towards caries [10].

The shedding of primary teeth does not usually cause any trouble. There is prolonged oozing from Gingival with the primary tooth very loosely attached to the soft tissue margin. In the rare case where it occurs, the constant disturbance of the loose crown by the tongue and the lip prevents the expected cessation of capillary bleeding by contraction and platelet plug, and the detachment of the offending tooth from the remaining attachment allows this mechanism to function satisfactorily [11-15].

## Dental treatment

An important problem in handling with hemophiliacs may be get acceptance of routine dental care, those patients with a family history of the diseases, and mainly if it involves some unfortunate dental incident, may have a great prejudice which must overcome and extended and repeated discussions may be needed make the parents realize how essential it is that their child should have dental care as a preventive measure.

The dental practitioner should, first of all, familiarize himself with the medical history of the patient. He needs particularly to know how severe the blood defect is since this will influence treatment choice [16].

A severely afflicted child should have an ambulance or other transport organized if the family has no private car or if the surgery's journey presents any real difficulty, especially if the public transport is a crowded bus. Such a vehicle is no place for these patients. He may arrive for dental treatment on some occasions in a wheelchair, with a leg in plaster or wearing calipers, and should have appropriate support when sitting in the dental chair. Apart from providing a proper leg rest if necessary, the dental chair is usually satisfactorily padded, but on some models, the additional seat for small patients has a rigid front edge, causing a hematoma on the leg's back if not padded further.

The further procedure with a new patient must be a full mouth examination and complete charting; notes must be made of the state of the Gingiva and oral hygiene, and the presence and extent of all caries and the malocclusion. The bite-wing co-operation of the child should be assessed after time has given to getting to know him [16,17].

## Prevention

It is an essential part of the dental care of hemophiliac patients. The attention must be directed to periodontal and caries aspects instruction in tooth brushing and repeated at intervals. The child and the parents should inform of the reason and the value of this rather tedious chore, and the lesson can be driven home by disclosing solutions or wafers. The toothbrush must not be hard, and it is better to use soft one more briskly. A battery-operated brush is a valuable aid, not only in easing the burden of bruising, but the small head allows the buccal surface of the maxillary molars and the lingual surface of the mandibular molars to appropriately cleaned without trauma or retching. The standard hand-operated toothbrush occasionally slips and causes abrasions, which may alarm the patient, a misfortune not possible with the battery brush. The latter has quite soft bristles and can use very effectively to give Gingival stimulation. Teeth should be brushed twice a day with a medium texture brush to remove plaque deposits. Dental floss or inter dental brushes should be used wherever possible. Toothpaste containing fluoride should be used in areas where natural fluoride is not present in the water supply. Fluoride supplements may also be prescribed if appropriate.

Discuss about a good diet with the parents is useful, emphasizing the need for fibrous food and avoiding between-meal carbohydrates snacks. Mention should be made of the importance of going to bed with clean teeth and no drinks or snacks other than water after tooth brushing.

If the patient's area of residence does not have a fluoridated water supply, fluoride tablets should be prescribed as a standard measure in prevention, indeed in all of these patients under the age of 8 to 10 years. The inquiry should be made pressed to have dental care. Symptoms of the defect do not often manifest themselves in infancy, and it is as well to encourage parents to give fluoride tablets to male infants

in hemophiliac families as a precautionary measure to protect the teeth as early as possible against dental caries, and hence against the possibility of the need for extractions in the years to come [16-18].

## Extractions

Cyklokapron (Tranexamic acid) or Epsilon amino caproic acid (EACA) is often used after dental procedures to reduce the need for replacement therapy. Oral antibiotics should only be prescribed if clinically necessary. Local hemostatic measures may also be used whenever possible following a dental extraction. Typical products include oxidized cellulose and fibrin glue. Prolonged bleeding and/or difficulty in speaking, swallowing, or breathing following dental manipulation should be reported to the hematologist/dental surgeon immediately. Non-steroidal anti-inflammatory drugs (NSAIDs) and aspirin must be avoided. An appropriate dose of paracetamol/acetaminophen every 6h for 2 - 3 days will help prevent pain following an extraction [19].

If extractions are unavoidable, these will be carried out in hospital as an inpatient by an experienced oral surgeon in conjunction with the hematologist and the physicians concerned. It is usually done under general anesthesia and with appropriate hematological preparation. A splint is made beforehand to protect the blood clot in the sockets, and if a large number of teeth require to be removed, it may be an advantage to limit the surgery to one jaw at a time, but this is a matter for consultation with the oral surgeon [20].

Although a technician is generally associated with a large hospital oral surgery department and is the usual person to construct the necessary splints, it may be more convenient in a particular case for the dental practitioner to undertake this. The construction of removable splint presents no problems. Good upper and lower alginate impressions of the mouth are needed, and are cast in hard plaster. The occlusion should be recorded so that the models can be articulated if necessary. The teeth to be extracted are cut off the models without any trimming of the Gingival margins so that the result represents the mouth after operation. an acrylic plate is made to cover the palate in the upper, or the lingual aspect of the lower jaw, as required, and is extended over the sockets of the extracted teeth to the buccal side. The object of this is to protect the sockets and their blood clots and not to provide pressure over them. The appliance is retained with Adams clasp cribs on appropriate molars supplemented by clasps on teeth further forward if necessary. It must be very adequately retained and stable where all the molars in the arch are being extracted, the retention is by the clasps in the anterior teeth, and if necessary, a buildup of the appliance over the distal parts of the arch to occlusal level in the manner of the bite block top occlude comfortably but not forcefully with the opposing teeth. This is worn for several days after operation and taken out only for cleaning, being replaced immediately [19-23].

## Periodontal problems

Apart from the gingivitis due to poor oral hygiene, this is not usually a problem in young children in the teenager there may be the same signs as in the normal patient. Scaling presents no real problem provided care is taken not to traumatize the gingiva unduly. Any bleeding which occurs is superficial and capillary in type, and will cease normally, such scaling must be meticulous since the smallest residuum of calculus and can cause gingival irritation with bleeding and pocket formation. Gingivectomy is contraindicated, however, as this involves more than the small end capillaries and may give rise to considerable persistent bleeding any small localized area of gingivitis should be investigated carefully and may be the result of a piece of calculus, a small foreign body, an overhanging filling or an area of traumatic occlusion.

Occasionally the gum flap over an erupting molar becomes inflamed and should be treated by painting under it toughly with 1 percent gentian violet or trichloroacetic acid and glycerin. Any acute oral infection should be dealt with promptly and effectively, and if there is any doubt as to the treatment, the advice of the physician in charge of the patient should be sought. Antibiotics should be given by the oral or intravenous route and never intra-muscularly [24-26].

## Conservation

The main difficulty in relation to conservation is the contraindication to local anesthesia deep injections for an inferior dental block and into the floor of the mouth should never be given even in a mild case damage to vessels frequently occurs in such injections and these areas have tissues spaces through which blood can track without difficulty. infiltration anesthesia should only be used with the agreement of the physician responsible who knows the hematological findings.

In cases of especial difficulty conservative procedures may be done under general anesthesia with the consent of the physicians who are responsible for the patients general care, this is done with endotracheal administration, the tube being passed by the mouth rather than the nose to avoid trauma to the adenoids. It necessitate admission to hospital for minimum of two or three days with stand by facilities for hemorrhage control in case of difficulties in passing the tube. Conservation under general anesthesia for these patients is not a form of treatment to be undertaken lightly [27,28].

Cavity preparation must be carefully carried out with good finger support to avoid any slip of instruments, and whenever possible prophylactic extension into susceptible areas should be done. If the tooth is particularly sensitive to instrumentation it is helpful to use diamond burs and to make the full extension at a depth just above the amelo dentinal junction at first. The cavity is then deepened to be required amount rapidly so that the painful part of the operation is very brief small burs are less painful than the larger sizes. Cavities involving the Gingival margin need present no real anxiety provided care is taken and cervical margin trimmers are used with caution any slight gingival bleeding ceases normally matrix bands should be used as usual, and indeed it is especially necessary to restore a good contact point and smooth junction interstitially with no overhang of the filling. Lack of care in finishing a class II filling can result in an inflamed inter dental papilla and recurrent bleeding problem. The matrix band, however, should be carefully placed and care taken to see what it is not pushed too far down and cause unnecessary damage to the periodontal fibers. A saliva ejector must be used with the caution. The control switch should be on a low power and it may be helpful to place a piece of gauze below the ejector end to protect the mucous membrane. A haematoma in the floor of the mouth is a serious matter and if it occurs, hospital advice must be sought.

Rubber dam clamps should not be used as they frequently cause marked crushing or bruising of the inter dental papilla and Gingival margin. if rubber dam is used, it should be retained with waxed floss. When polishing the filling it is wise to avoid the use of bristle brushes as these can cause abrasion to the adjacent soft tissue and are subject to slip. Finishing burs and rubber cups should be satisfactory when used with the appropriate pastes [28].

## Pulp treatment

In permanent teeth vital exposures may be capped if the operator has success with this type of treatment. Pulpotomy presents a problem of anesthesia but may be carried out with ordinary inhalation anaesthesia if this is available with a component anaesthetist. A mouth prop should be inserted before the start of induction as it is less traumatic than the opening the mouth with a gag at a later stage. There is said to be no hemorrhage problem of the pulp associated with this type of operation, but it seems possible that pressure due to bleeding could build up in the sealed chamber and so produce pulp death. Where extirpation of the vital pulp is indicated, this is the best carried out with the aid of devitalizing paste since local anesthesia is contraindicated. Following this, standard root canal procedures can be used.

In primary teeth, the exposure of a vital pulp should be treated by revitalization rather than attempting pulp capping. Incisors are root filled standard techniques, being very careful not to penetrate the apex and cause damage to the permanent successor. The root filling material must be resorbable to allow normal shedding. Primary molars on the hand, do not lend themselves to root filling, and a techniques of sterilization and mummification must be substituted. Such treatment must be followed periodically by radio graphic check of the apical condition [29,30].

### Malocclusion

Any plan of treatment for malocclusion in a hemophiliac patient must take into account the severity of the bleeding problem. The need for extractions to relieve crowding, the attitude of the child to the malocclusion and the degree of the blood defect must all be discussed with the hematologist and the physician in charge of the patient. Normally extractions will only be considered in the less severe haemophiliac, but in the more severe case a possible consideration is whether a tooth may cause trauma to the soft tissues. An example of this is a maxillary canine crowded out of the arch buccally to the extent of being a constant threat of trauma to the lip.

Appliance therapy should be minimal for the severely afflicted child but some malocclusion are more harmful if left. a particular example is the in standing maxillary incisor in traumatic occlusion. Such a situation leads to periodontal pocketing on the mandibular incisors if not treated, and the simple appliance will usually correct it quickly. Removable appliances are used with Adams or similar clasps without free ends which may cause trauma if inadvertently bent out of the place, and well adjusted. Andersen or monobloc appliance is suitable in appropriate cases, in milder hemophiliacs more elaborate orthodontic treatment can be done and fixed appliances may be used if great care is taken [31-33].

For procedures that do require increment in the factor levels, there may be four therapeutic management options depending on the type of hemophilia, namely:

1. Coagulation factor replacement therapy
2. Release of endogenous factor stores using desmopressin (DDAVP)
3. Improving clot stability by antifibrinolytic drugs, for example Tranexamic acid, to reduce the need for replacement therapy
4. Local hemostatic measures (such as suturing, and local measures, such as the use of oxidized cellulose) [33].

For example, infiltration, intra-papillary, and intra-ligamentary injections are often done under factor cover (20 - 40%), though it may be possible for those with adequate experience to administer these injections without it. It is advisable that complicated dental procedures, such as dental extraction or surgical procedures carried out within the oral cavity, should be performed in a Hemophilia Treatment Center [34].

## Conclusion

It is not uncommon for dental practitioners to encounter patients with bleeding disorders in their daily practice; therefore, it is essential to be able to identify such patients and safely manage their dental treatment. Close liaison between the dental surgeon and the hemophilia team is essential to provide good comprehensive dental care.

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