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Orthodontic Care for Hemophilic Patients

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Abstract

Objective: This review is an attempt to provide guidelines for orthodontic evaluation and treatment of individuals with Hemophilia. **Results:** The literature data allowed us to observe that hemophilia nowadays is not an absolute contraindication to orthodontic treatment. The coordination and communication of haematologist and professionals in the oral cavity are essential. **Conclusion:** Thanks to advances in orthodontics, there is no contraindication and the decision to carry out treatment is made with the same criteria as any other patient.

Subject Areas

Dentistry

Keywords

Hemophilia, Bleeding Disorders, Orthodontic Treatment

1. Introduction

The dental management of patients with haemophilia has been an object of interest to dental professionals in recent decades. The orthodontic treatment of these patients has been neglected for years for fear of bleeding, which involved complications in the occlusal, dental and periodontal state of the same [1] [2]. Orthodontic treatment is an elective procedure for essentially all patients, and certainly for those with chronic medical problems. There is clear evidence that correction of disfiguring dental problems contributes in an important way to an individual's self-esteem and affects social integration and interaction [2] [3] [4].

This review is an attempt to provide guidelines for orthodontic evaluation and treatment of individuals with Hemophilia.

2. Diagnosis

Hemophilic patients must be thought of as special patients. It is of vital impor-

tance to know the type and severity of the disease, the presence or not of inhibitor agent, the current medications, family history of the condition, previous hospitalizations, transfusions, existence of other diseases and previous dental treatments [1].

Approximately 80 percent of affected persons have classic hemophilia, otherwise known as hemophilia A, in which antihemophilic blood plasma factor VIII is missing. Another 15 per cent have Christmas disease, or hemophilia B, in which a different protein, factor IX, is absent from the blood. Hemophilia C, is the result of a deficiency of PTA (Plasma Thromboplastin Antecedent), otherwise known as factor XI. This is a milder form of the disease and is considered to be rarer because it is not diagnosed as frequently. Finally, the fourth relatively common form of hemophilia, Von Willebrand's disease, also hereditary, affects both males and females, and is due to a deficiency of factor VIII as well as an absence of the cofactor which makes platelets sticky [4] [5] [6] [7].

2.1. Biological Background

The blood coagulation mechanism does not function normally in hemophilia. The clotting process involves a series of biochemical reactions with each initiated sequentially by the preceding reaction. If any one of the clotting factors is deficient, the time required for the formation of a clot is greatly prolonged [3] [5].

The diagnostic laboratory findings in hemophilia can be summarized as follows: a prolonged activated partial thromboplastin time (APTT), normal prothrombin time (PT), normal bleeding time (BT) [5] [8].

The severity of haemophilia depends on the amount of this coagulation factor. The normal concentrations of clotting factor are between 50% and 150% of average value and the minimum level of a factor for adequate haemostasis is 25%. Haemophilia is classified as mild, when the decrease of factor is 5% - 25% of normal; moderate, when the decrease is 1% - 5%; and severe when the decrease is less than 1%. When levels of clotting factor are found below 1% it can cause spontaneous bleeding [1].

2.2. Genetic Background

Hemophilia A and hemophilia B are sex-linked diseases transmitted to the child by a gene on one of his mother's two X chromosomes [1] [9]. Thereby, sons of carriers have a 50:50 chance of developing hemophilia while daughters of carriers have a 50:50 chance of being carriers. All daughters of an affected male are carriers but sons are normal [5].

2.3. Oral Manifestations

Hemophilia is characterized by bleeding from multiple sites, frequently manifested in the mouth as gingival and post-extraction haemorrhages. Patients may also have multiple oral bleeding events over their lifetime, depending on the severity of hemophilia. Bleeding episodes are more frequently encountered in se-

vere hemophilia, followed by moderate hemophilia, and are minimal in mild hemophilia. Poor oral hygiene and iatrogenic factors can also induce the oral bleeding. Persistent oral bleeding is seen in about 14% of all hemophilic patients. In toddlers, oral ulcerations and ecchymosis involving lips and tongue are common [6] [10].

3. Orthodontic Treatment Considerations in Hemophilic Patients

3.1. Interceptive Orthodontic Treatment

As the prevalence of malocclusion in children with hemophilia is similar to the rest of the population and the long-term outlook is good, orthodontic treatment is often requested [3] [11]. Orthodontic treatment does not pose a risk for the health of these children, on the contrary, it affects the general health of the child more positively. Secondly, early diagnosis of developing malocclusion can reduce complex orthodontic problems [12] [13]. Therefore, preventive and interceptive orthodontic treatments performed in the early period are of great importance for these patients. Paediatric dentists and orthodontists should be educated in this matter [1]. The orthodontist should carefully weigh the advantages of functional appliances in these children. Intense orthodontic or orthopaedic forces such as extra oral anchorage or maxillary distraction should be managed with more care [13].

Functional appliances that reposition the mandible may produce bleeding both intraorally from tissue irritation and within the temporomandibular joints, making this sort of therapy less desirable for these patients than might be thought. Prolonged orthodontic treatment increases the risks of a bleeding episode, and therefore, orthodontic treatment in conjunction with planned orthognathic surgery with prior transfusion should be a major consideration for them. For this reason, orthognathic surgery to correct a severe jaw discrepancy may be a better choice for a hemophiliac patient than the apparently more conservative but prolonged orthodontic growth-modification approach [10]. As with any invasive procedure, orthognathic surgery requires preparation of the patient with transfusion to replace the missing clotting factors, but with proper precautions, this surgery is entirely feasible [14]. The literature presents only one study about the parent education, after asking "Do you think orthodontic treatment would affect your child's health negatively?" question, the majority of the parents are not educated enough in this matter as 37.93% replied they do not know; while 34.48% think it would. For this reason, it is important to inform the family early and to initiate preventive treatment especially in young children at an earlier age. Parents need to be informed more about the possibility that haemophiliac children can have orthodontic treatment [12].

3.2. The Risks of Orthodontic Treatment

Patients with haemophilia and related bleeding disorders require special consid-

eration in two areas: The viral infection risk and the bleeding risk [2] [12] [13].

3.2.1. The Viral Infection Risk

The usual treatment for patients with hemophilia is through the use of intravenous replacement of the missing or deficient coagulation factor. This can be done either prophylactically or at the time of injury, bleeding, or surgical procedure. Factor concentrates are derived from human blood donations. Since the mid-1980s methods of manufacture have been developed to remove hepatitis B, C, and HIV from human derived concentrates. However, the continued use of concentrates, despite careful donor selection and screening, and improved methods of manufacture, still carries a small risk of transmitting serious transfusion derived viral infection. Patients with hemophilia usually have received multiple transfusions of blood and blood products, which leads to a greatly increased incidence of hepatitis. They should be considered carriers for hepatitis until proved otherwise [11] [12] [15] [16]. Special precautions should be taken in handling these patients to prevent the possible spread of hepatitis virus to the dentist, staff members, or other patients. These precautions include measures that prevent exposure to serum or saliva such as the use of protective eyewear, gloves, face masks, and autoclave or formaldehyde vapour pressure sterilization of all instruments. The use of recombinant DNA technology factor has eliminated the risk of blood-borne pathogen infections, such as HIV or hepatitis, which is possible with human-derived products. Another therapeutic intervention provides antifibrinolytic effect and stabilizes clots by preventing clot break down, such medications include tranexamic acid and Amicar [8].

3.2.2. The Bleeding Risk

Orthodontic treatment is certainly not contraindicated, but caution should be exercised to avoid any lacerations during procedures. Direct bonding of attachments rather than fitting bands helps to reduce these risks. Care should be taken to ensure that there are no sharp edges or wires protruding from the orthodontic appliances. Currently, this does not happen because the bands are prefabricated and can be replaced by tubes and the technique is not multiband but multibrackets, bonded directly or indirectly, thereby minimizing the possible damage to the gums. The attitude has changed and patients with haematological disorders are now treated in the same way as the normal orthodontic patients [1] [9] [11] [12] [17]. Overall, all devices that are used in hemophilic patient's orthodontic treatment naturally should be had at least potential to cause injury. In these patients, fixed appliances are preferred because removable appliances will cause gingival irritation [2] [18]. "Self-ligating" brackets compared to "conventional" brackets are more appropriate. The orthodontist should be very careful when placing arch wires because they contain sharp end, and make sure that end wire is completely cut-off. Overall duration of treatment should be reduced to a minimum so that was less complex problems [1]. To assess the risk of orthodontic treatment is convenient to divide the treatment procedures into two groups:

Invasive procedures that can generate bleeding: Extractions, surgical manipulation of periodontal tissues, including an exhibition of a tooth, placement of bands with a subgingival extension and orthognathic surgery, and non-invasive procedures with minimal risk of bleeding: making impressions, placement of fixed appliances with direct bonding, stripping, routine adjustment of equipment and insertion and adjustment of removable appliances. If orthodontic treatment is in the first group, the patient will require a preoperative evaluation and postoperative follow-up. This is not necessary for those patients who are in the second group of the classification [1] [2]. Certainly, any intraoral use of dental instruments or materials provides the potential for a laceration or puncture wound and so may inadvertently become invasive, but, for the most part, orthodontic appliance therapy is non invasive. During orthodontic treatment with fixed appliances, oral hygiene is particularly important to avoid inflamed and oedematous gingival tissues, which are prone to hemorrhage [9] [17] [19].

3.3. Tooth Extraction and Orthognathic Surgery in the Orthodontic Treatment

In general, orthodontic treatment is not contraindicated in patients with bleeding disorders [1] [3]. If tooth extraction or other surgery is required in patients with severe bleeding disorders, they are usually hospitalized and given transfusion of the missing clotting factor in advance of the procedure, with serum preparations that supply the missing clotting factors [6]. The indications for invasive procedures must be carefully evaluated, but such procedures are not contraindicated if effective replacement therapy can be provided. But wherever possible, a non-surgical approach should be adopted [2] [12]. It is well established that routine orthognathic surgery procedures have minimal bleeding when soft tissues are reflected properly and the vessels are appropriately protected [8]. Because orthognathic procedures are performed in close anatomic proximity to multiple arterial and venous systems, a protocol must be available to achieve adequate intraoperative hemostasis. Such complications in patients with bleeding disorders can be fatal, and it is therefore necessary that appropriate preparations take place [8]. Management of patients with hereditary bleeding disorders mandates a close cooperation between haematologists and surgeons. It is therefore imperative that a plan be generated to decrease chances of unexpected intraoperative and postoperative hemorrhage, and thorough preparation must be established in case such hemorrhage occurs [8] [17]. According to a study, it is better all teeth that to be extracted for orthodontic treatment was removed in one session. While in another study stated that it was better the teeth that to be extracted for orthodontic treatment, reduced to a minimum. Laboratory investigations are necessary before any dental procedures that are likely to cause postoperative bleeding. These include the activated partial thromboplastin time (aPTT), bleeding time, and factor coagulant level in plasma [17]. Although local haemostatic measures often provide successful results in patients with bleeding

disorders, these measures are insufficient to control postoperative bleeding in some patients with severe haemophilia [6]. To help prevent and manage post-operative bleeding episodes, the dentist should consult with the haematologist preoperatively by sending a referral letter that explains the intended dental procedures, underlines the medical conditions, and describes the types of post-operative local haemostatic measures that will be applied [20.]

Postoperatively, it is recommended that the patient avoid any analgesic medications that increase the possibility of bleeding, such as aspirin and other non-steroidal anti-inflammatory drugs. Paracetamol can be used safely. Some dental procedures may necessitate antibiotic use; therefore, the haematologist should be consulted [3] [21].

4. Conclusions

The literature data allowed us to observe that hemophilia nowadays is not an absolute contraindication to orthodontic treatment. The coordination and communication of haematologist and professionals in the oral cavity are essential.

Special orthodontic considerations can be summarized as follows:

1) Maintaining and establishing good oral hygiene will prevent gingival bleeding;

2) Self-ligating brackets are preferable to conventional brackets;

3) The archwires should be secured with elastomeric modules rather than ligature wire, later might have a risk of laceration of mucosal surface;

4) Fixed appliances are preferable to removable appliances as the latter can cause gingival irritation;

5) The duration of orthodontic treatment should be kept to a minimum to reduce the potential for complications.

Conflicts of Interest

The authors declare no conflicts of interest.

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Abbreviations

PTA = Plasma Thromboplastin Antecedent
APTT = Activated Partial Thromboplastin Time,
PT = Prothrombin Time
BT = Bleeding Time
HIV = Human Immunodeficiency Virus,
DNA = Deoxyribonucleic Acid
aPTT = Activated Partial Thromboplastin Time
NSAIDs = Non-steroidal anti-inflammatory drugs.