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Specific Dental Management of Hemophilia A and B, von Willebrand Disease and Acquired Hemophilia

Hemophilia A

Hemophilia A is a congenital bleeding disorder, X-linked, characterized by a quantitative deficiency of coagulation factor VIII. From a dental standpoint, this condition requires strict interdisciplinary coordination and prior consultation with the treating hematologist.

Prevention:

Since any bleeding event may represent a medical emergency, the dental approach should be focused on prevention. Personalized oral hygiene protocols, intensive plaque control, topical fluoride application, and regular

professional follow-ups should be implemented from early ages. Dental prophylaxis should be performed with minimally traumatic techniques, and ultrasonic devices should be avoided unless anti-hemorrhagic prophylaxis has been provided.

Dental Caries:

Restorative procedures must be carefully planned. If deep cavities are involved, infiltrative anesthesia is recommended under factor VIII concentrates coverage. In selected cases, performing multiple restorations in a single appointment may be feasible if full hemostatic support is ensured.

Periodontal Disease:

Non-surgical periodontal therapy may be performed under appropriate hematologic coverage and local bleeding control. Scaling and root planning should be carried out quadrant by quadrant, using delicate instrumentation and avoiding sharp curettes in the absence of systemic factor replacement. Periodontal surgeries should be avoided unless performed in hospital settings under full hematological supervision.

Dental Extractions:

Any surgical indication requires prior hematologic evaluation. Factor VIII must be administered about 15-30 minutes before the procedure and maintained for 48–72 hours postoperatively, according to the hematologist's protocol. Nerve block anesthesia should be avoided without full coverage. Use of absorbable sutures, local compression, and topical antifibrinolytics such as tranexamic acid is strongly advised.

Hemophilia B

Hemophilia B is caused by factor IX deficiency. Although its clinical presentation is similar to hemophilia A, the pharmacokinetics of factor IX necessitate a different treatment protocol.

Preventive and Conservative Management:

Oral disease prevention is fundamental. Reinforcement of oral hygiene, professional plaque control, fluoride therapy, and regular follow-ups are essential. No invasive procedures should be performed without hematology clearance.

Periodontal Disease and Dental Caries:

Non-surgical periodontal therapy may be carried out under systemic hemostatic coverage if spontaneous gingival bleeding or moderate to severe periodontitis is present. Atraumatic restorative techniques (ART) are preferred for patients without hematologic coverage. High-speed instrumentation and rubber dam isolation should only be used in patients with adequate levels of factor IX.

Surgical Procedures:

Any surgical dental intervention requires coordination with hematology. Factor IX should be administered according to individual needs. Given its longer half-life, if compared to the Factor VIII concentrates, extended monitoring is often necessary. Extractions must be minimally traumatic, using conservative techniques, local antifibrinolytics, and anticipation of delayed bleeding in multi-rooted teeth.

von Willebrand Disease

Von Willebrand Disease (vWD) is the most common inherited bleeding disorder. Its clinical severity varies depending on the type (1, 2, or 3), affecting both the quantity and/or functionality of von Willebrand Factor (vWF), and frequently involving secondary factor VIII deficiency.

Preoperative Evaluation:

It is essential to obtain a full hematological report prior to any dental intervention. The decision to use desmopressin (DDAVP), vWF only, or vWF and FVIII concentrate must be made exclusively by the hematologist.

Oral Disease Prevention:

Prevention is the cornerstone of care. Soft toothbrushes, fluoride toothpaste and mouthrinses, and chlorhexidine-based antiseptics are strongly recommended. Professional monitoring should be frequent to prevent gingivitis and spontaneous bleeding.

Caries and Periodontal Treatment:

Restorative care should be minimally invasive. Infiltrative anesthesia may be used, but nerve blocks must be avoided unless there is hemostatic coverage. Vasoconstrictors may enhance local bleeding control.

Scaling and Periodontal Surgery:

Periodontal surgery is not advised unless under hematologic supervision. Supragingival scaling may be done cautiously. Subgingival or invasive procedures require prior administration of systemic agents and use of topical antifibrinolytics like tranexamic acid.

Extractions

All extractions must be planned in conjunction with hematology. DDAVP or vWF with or without FVIII concentrates should be administered according to the type and severity of vWD. Conservative surgical technique, use of resorbable sutures, compression, and tranexamic acid rinses during the first 48 hours post-op are strongly indicated.

Acquired Hemophilia

Acquired hemophilia is a rare autoimmune bleeding disorder characterized by the development of autoantibodies (inhibitors) against coagulation factor VIII. It primarily affects older adults and may be associated with autoimmune diseases, malignancies; in other cases, it can appear during pregnancy or the

postpartum period, or may affect people idiopathically. Unlike congenital hemophilia, bleeding episodes are often spontaneous, severe, and unrelated to trauma, but sometimes the diagnosis is made after post-surgical hemorrhages or large hematomas at the sites of blood sampling.

General Dental Considerations:

All dental procedures must be preceded by a comprehensive hematologic evaluation. In most cases, standard factor VIII replacement therapy is ineffective due to the presence of circulating inhibitors. Therefore, hemostatic management relies on bypassing agents such as recombinant activated factor VII (rFVIIa), activated prothrombin complex concentrates (aPCC), or recombinant porcine factor VIII concentrate, which must be administered strictly under the supervision of a hematologist. In any case, it is always advisable to postpone dental procedures until the inhibitor has been eliminated, unless they are urgent.

Preventive Approach:

In cases of prolonged persistence of the antifactor VIII inhibitor or relapse of the inhibitor after effective immunosuppressive treatment, dental care must be centered on strict prevention of oral diseases, due to the high and unpredictable bleeding risk. Oral hygiene instructions should be reinforced with the use of soft-bristled toothbrushes, fluoride toothpaste, 0.12% chlorhexidine rinses, and frequent clinical monitoring. Dental cleanings should be performed using non-invasive, manual techniques, avoiding ultrasonic scalers.

Restorative and Periodontal Therapy:

Only minimally invasive treatments should be considered, and only in patients previously stabilized hematologically. Manual instrumentation should be preferred over ultrasonic devices. Non-surgical periodontal therapy must be performed with extreme caution and only under hematologic coverage if there is evidence of gingival bleeding. Periodontal surgery is contraindicated in outpatient dental settings.

Oral Surgery and Extractions:

Tooth extractions and other invasive procedures must be performed in hospital-based settings with full hematologic support. The use of bypassing agents, or recombinant porcine factor VIII concentrate is mandatory. Additional local measures, such as resorbable hemostatic sutures, pressure with surgical gauze, and tranexamic acid mouth rinses for 48–72 hours, are critical. The risk of delayed or recurrent bleeding must be anticipated and the patient monitored closely.

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