JOURNAL OF HEALTHCARE SCIENCES
Volume 4 Issue 11 2024, Article ID: JOHS2024000929
http://dx.doi.org/10.52533/JOHS.2024.41107
e-ISSN: 1658-8967

Review

Dental Care for Children with Hemophilia Managing Bleeding Risks and Oral Health

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Received: 16 October 2024, Reviewed: 22 November 2024, Accepted: 24 November 2024, Published: 25 November 2024.

Abstract

Hemophilia is a genetic bleeding disorder characterized by a deficiency in clotting factors, primarily factor VIII or IX, which leads to impaired coagulation and prolonged bleeding episodes. This condition presents significant challenges in dental care, especially for children. Hemophilic patients are prone to spontaneous bleeding in the oral cavity, particularly from the gingiva, due to the high vascularization of oral tissues and the frequent exposure to mechanical trauma. Common oral manifestations include gingival bleeding, periodontal disease, and dental caries, which are exacerbated by difficulties in maintaining proper oral hygiene. The pathophysiology of hemophilia disrupts the normal coagulation cascade, making even minor dental procedures, such as cleanings or fillings, risky without adequate preparation. Prophylactic treatment with clotting factor replacement therapy is often necessary before invasive procedures to reduce the risk of excessive bleeding. Additionally, the use of antifibrinolytic agents and local hemostatic measures, such as sutures or fibrin sealants, helps control bleeding during dental interventions. Children with hemophilia also face an increased prevalence of dental caries and periodontal disease due to the challenges in maintaining oral hygiene and the frequent consumption of sugary medications. These factors lead to a higher incidence of dental issues that require special management strategies. Preventive care, including regular dental check-ups, fluoride treatments, and minimally invasive procedures, is critical to reducing the need for more complex interventions. Successful management of dental care in hemophilic patients requires a multidisciplinary approach involving collaboration between dentists and hematologists. Individualized care plans tailored to the severity of the condition and the type of dental procedure are essential to minimize bleeding risks and ensure safe and effective treatment. Advances in dental technologies and treatment approaches continue to improve the outcomes and quality of life for children with hemophilia.

Keywords: *Hemophilia, dental care, bleeding risks, oral health, pediatric dentistry*

Introduction

Hemophilia is a group of inherited bleeding disorders characterized by a deficiency or dysfunction of clotting factors, primarily factor VIII (hemophilia A) and factor IX (hemophilia B). This deficiency impairs the blood clotting process, leading to prolonged bleeding after injury, surgery, or dental procedures. Hemophilia is typically classified as mild, moderate, or severe, depending on the level of clotting factor present in the bloodstream. Severe forms of the condition are often associated with spontaneous bleeding episodes, particularly affecting joints and muscles (1). In children with hemophilia, dental care presents unique challenges due to the elevated risk of bleeding and the need for careful management to prevent complications.

The oral health of children with hemophilia can be significantly impacted by their condition. The underlying pathophysiology of hemophilia, which involves a disruption in the coagulation cascade, means that even minor oral traumas can result in prolonged or severe bleeding (2). Moreover, these children are at increased risk for oral health issues such as gingivitis, periodontal disease, and dental caries. Poor oral hygiene and delayed or inadequate dental treatment, exacerbated by fears of bleeding complications, contribute to the higher prevalence of oral health problems in this population (3). Early and regular preventive dental care is essential for minimizing these risks and ensuring overall well-being.

Management of dental procedures in children with hemophilia requires a multidisciplinary approach, involving coordination between hematologists, pediatric dentists, and other healthcare professionals. Effective management strategies include administering clotting factor replacement therapy before invasive procedures and employing local hemostatic measures such as tranexamic acid or fibrin sealants (4, 5). Additionally, dentists must be cautious when performing routine treatments such as extractions, restorative procedures, or periodontal care, as these interventions may trigger significant bleeding without proper precautions.

The most common dental issues among children with hemophilia include bleeding gums, recurrent oral ulcerations, and dental caries, often requiring special attention from dental professionals. Managing these issues requires understanding the interplay between the disease's pathophysiology and oral health, emphasizing the importance of preventive care, regular check-ups, and the judicious use of local hemostatic agents during any necessary dental interventions (2). This review explores the pathophysiology of hemophilia, its impact on oral health, strategies for managing bleeding risks during dental procedures, and common dental problems observed in children with hemophilia.

Review

Children with hemophilia face unique challenges in maintaining oral health due to the nature of their bleeding disorder. The pathophysiology of hemophilia, characterized by deficient clotting factors, results in a higher risk of prolonged bleeding following minor trauma or routine dental procedures. This necessitates a cautious approach when managing their dental care. Preventive strategies, such as regular dental check-ups and meticulous oral hygiene practices, are critical for reducing the risk of bleeding and other oral health issues (6, 7). Additionally, collaboration between dentists and hematologists ensures that appropriate pre-treatment interventions, like clotting factor replacement, are administered when necessary to reduce bleeding risks during invasive procedures.

Common oral health problems in children with hemophilia include gingival bleeding, periodontal disease, and dental caries. Gingival inflammation, exacerbated by poor oral hygiene, often results in spontaneous bleeding, which can complicate dental care (2). Furthermore, dental caries and periodontal disease are more prevalent due to both the difficulty in maintaining proper oral hygiene and the hesitation to seek treatment due to concerns about bleeding. These factors highlight the importance of preventive dental care and the use of localized hemostatic measures, such as antifibrinolytics and

http://dx.doi.org/10.52533/JOHS.2024.41107

topical agents, to manage bleeding effectively during dental procedures.

Pathophysiology of Hemophilia and Its Implications for Dental Care

Hemophilia is a genetic disorder characterized by a deficiency in clotting factors, most commonly factor VIII in hemophilia A and factor IX in hemophilia B. These factors are essential components of the coagulation cascade, a series of reactions that lead to the formation of a stable blood clot at the site of injury. In individuals with hemophilia, the absence or reduced function of these factors impairs the body's ability to form a clot, leading to prolonged bleeding episodes even after minor injuries. The severity of hemophilia is classified based on the percentage of clotting factor activity in the blood, with severe cases having less than 1% activity, moderate cases between 1-5%, and mild cases between 6-30% (8). This deficiency profoundly affects not only systemic health but also oral health, necessitating tailored dental care to prevent and manage bleeding complications.

The bleeding diathesis associated with hemophilia significantly complicates dental care, especially in children. Routine oral traumas, such as those caused by brushing or flossing, can lead to prolonged gingival bleeding. Additionally, any invasive dental procedure, including simple extractions, restorations, or cleanings, poses a risk for excessive bleeding that can be difficult to control without preemptive management. Dental practitioners must fully understand the pathophysiology of hemophilia to assess bleeding risks accurately and develop appropriate preventive and treatment strategies for affected patients (9).

In hemophilia, the coagulation cascade is disrupted at the point where factor VIII or IX is required for the activation of factor X. This step is crucial for converting prothrombin to thrombin, the enzyme responsible for the transformation of fibrinogen into fibrin, which forms the structural matrix of a blood clot. Without adequate levels of factor VIII or IX, the activation of factor X is significantly delayed or incomplete, resulting in a weak or absent clot (10). As a result, children with hemophilia are highly susceptible to prolonged bleeding episodes, both spontaneously and in response to minor injuries, including those sustained during everyday oral hygiene practices. The oral cavity, with its rich blood supply, becomes a high-risk area for significant blood loss, especially when gingival tissues are inflamed or damaged. This pathophysiological framework extends to the heightened risk of spontaneous bleeds, particularly in the soft tissues of the mouth. Hemophilic children are often observed to develop oral hematomas or spontaneous gum bleeding due to even minor irritations, such as food impaction or accidental bites (11). These episodes can be particularly distressing for children and their caregivers and may discourage consistent oral hygiene practices, further compounding the risk of periodontal disease and dental caries. Additionally, the chronic inflammation caused by recurrent bleeding episodes creates an environment that facilitates the progression of oral diseases, leading to a vicious cycle of oral health deterioration.

The implications of hemophilia for dental care extend beyond routine hygiene practices to more invasive procedures, such as extractions, fillings, or even orthodontic interventions. Each procedure carries a unique set of risks due to the fragile hemostatic balance in hemophilic patients. For instance, tooth extractions are among the most common dental procedures that can trigger significant post-operative bleeding. In hemophilic children, these extractions may lead to uncontrolled bleeding if appropriate pre-procedure measures are not taken, such as the administration of clotting factor replacement therapy or antifibrinolytic agents like tranexamic acid (12). The role of the dentist in these cases goes beyond mere technical skill; it requires close collaboration with the patient's hematologist to ensure that clotting factor levels are optimized before any invasive procedure.

Moreover, the prolonged bleeding time observed in hemophilia patients can lead to complications such as infection or delayed healing after dental procedures. The lack of efficient clot formation means that wounds remain open for longer periods, providing an entry point for bacteria and increasing

http://dx.doi.org/10.52533/JOHS.2024.41107

the risk of post-operative infections. Dentists must adopt strategies that promote rapid healing and minimize bacterial colonization, such as using minimally invasive techniques, suturing, and prescribing antibiotics when necessary. Furthermore, the use of hemostatic agents, like fibrin glue or gelatin sponges, can aid in controlling local bleeding and promote faster wound closure (9). Given the delicate balance between preventing bleeding and managing oral health in children with hemophilia, dental professionals must be equipped with knowledge of the underlying pathophysiology and how it impacts their patients' care. Understanding these mechanisms allows for better risk assessment and planning, helping to ensure that both routine care and more complex dental procedures are carried out safely.

Pathogenesis and Common Oral Manifestations in Hemophilic Patients

The pathogenesis of oral manifestations in hemophilic patients is closely tied to the underlying deficiency of clotting factors that characterize the disorder. Hemophilia, whether type A (factor VIII deficiency) or type B (factor IX deficiency), disrupts the coagulation cascade, leading to impaired blood clot formation and extended bleeding times. This not only affects systemic health but also has direct consequences for oral tissues, where even minor injuries or routine activities like toothbrushing can result in prolonged or spontaneous bleeding (13). The high vascularization of the oral mucosa, combined with frequent exposure to mechanical forces, makes the mouth particularly vulnerable in hemophilic patients, resulting in a range of oral health complications that require vigilant management.

One of the most common oral manifestations in hemophilic patients is gingival bleeding. This can occur spontaneously or because of mild trauma, such as during tooth brushing or flossing. Gingival tissues are rich in blood vessels, and even slight irritation or inflammation, such as that caused by gingivitis, can lead to significant bleeding episodes in individuals with hemophilia. This recurrent bleeding not only poses a direct health risk but also contributes to the progression of periodontal disease by allowing bacterial colonization in areas of chronic inflammation (3). Gingival bleeding in hemophilic patients often becomes a chronic issue, further complicated by inadequate oral hygiene practices due to fear of triggering bleeding episodes.

Another common oral manifestation in these patients is hemophilic pseudotumors, although these are rare in comparison to other manifestations. Pseudotumors develop because of repeated hemorrhages into soft tissues, including the jaw, leading to progressive bone destruction and swelling. These masses are usually non-malignant but can cause considerable discomfort and require surgical intervention if they interfere with oral functions, such as chewing or speaking. In hemophilic patients, pseudotumors in the oral cavity or jaw can lead to complications, particularly when they necessitate invasive procedures that carry a high risk of bleeding (14). Early detection and management of such conditions are crucial to prevent more severe outcomes and minimize the risk of prolonged bleeding.

Oral mucosal hemorrhages are also frequently observed in hemophilic patients. These hemorrhages, which are often present as small red or purplish spots (petechiae) or larger areas of bruising (ecchymosis), are caused by the extravasation of blood into the mucosal tissues due to capillary fragility and the lack of clot stabilization. These lesions are not only painful but may also complicate oral hygiene and feeding in younger patients. Moreover, mucosal hemorrhages increase the risk of secondary infections, particularly fungal infections such as oral candidiasis, which can exacerbate discomfort and further compromise oral health (15). The presence of these hemorrhages is a clear indicator of the systemic bleeding disorder, and their management typically requires both local measures and systemic treatments to restore clotting factor levels.

In addition to these direct bleeding-related complications, dental caries is another concern in hemophilic patients. Fear of bleeding often leads to neglected oral hygiene, which, in turn, increases the risk of caries. Furthermore, children with

http://dx.doi.org/10.52533/JOHS.2024.41107

hemophilia may experience difficulties in maintaining a consistent oral hygiene routine due to the discomfort associated with bleeding gums and mucosal lesions. This increased risk of dental caries, combined with the challenge of performing restorative dental procedures in patients with compromised hemostasis, complicates their overall oral care. The management of caries in hemophilic patients requires careful planning, including pre-treatment with clotting factor concentrates and the use of minimally invasive techniques to reduce the risk of post-procedural bleeding (16).

Dental Management Strategies for Minimizing Bleeding Risks

The dental management of children with hemophilia requires careful planning and an understanding of both the condition's pathophysiology and the interventions needed to minimize bleeding risks during routine and invasive procedures. Dentists must adopt a multidisciplinary approach, working closely with hematologists to ensure that patients receive appropriate pre-procedural treatments to mitigate the risk of excessive bleeding. The cornerstone of managing these patients lies in individualized care plans that consider the severity of hemophilia and the type of dental procedure being performed.

One of the primary strategies for reducing bleeding risks is the administration of clotting factor concentrates before any invasive dental procedure. For patients with moderate to severe hemophilia, prophylactic infusions of factors VIII or IX can significantly reduce the likelihood of post-operative bleeding. These infusions are usually administered within an hour prior to the dental procedure to ensure that clotting factor levels are sufficiently elevated during the intervention (2). The dosage and timing of these infusions are determined by the patient's hematologist, based on the severity of the hemophilia and the nature of the procedure. For minor procedures like cleanings or fillings, a single dose may suffice, while more complex procedures, such as extractions, may require additional doses post-operatively to maintain adequate clotting factor levels.

The use of antifibrinolytic agents such as tranexamic acid plays a crucial role in minimizing bleeding during and after dental procedures. Tranexamic acid works by inhibiting the breakdown of blood clots, thereby enhancing clot stability and reducing the risk of prolonged bleeding. It is commonly used both systemically, in the form of oral or intravenous administration, and topically, applied directly to the surgical site. Studies have shown that the combination of clotting factor replacement and tranexamic acid significantly reduces bleeding risks in hemophilic patients undergoing dental surgery (17). This combination allows for safer management of even invasive procedures, such as tooth extractions or periodontal surgeries, which would otherwise carry a high risk of complications.

Local hemostatic measures are also a key component of dental management in hemophilia patients. The use of absorbable hemostatic agents, such as oxidized cellulose or fibrin sealants, can be applied to surgical sites to promote clot formation and prevent excessive bleeding. These agents provide mechanical support to the clot and help seal the blood vessels, reducing the risk of post-operative bleeding. Suturing is another important technique for controlling bleeding, especially after extractions or other invasive procedures. Careful, tension-free suturing can help stabilize the soft tissues and encourage faster healing while minimizing the likelihood of re-bleeding (18). Dentists should be prepared to use a combination of these local measures, particularly in procedures where bleeding is anticipated to be a significant risk.

For non-invasive procedures, such as routine dental cleanings or restorative work, minimizing trauma to the tissues is critical. The use of atraumatic techniques, such as laser dentistry or air abrasion, can reduce the risk of gingival bleeding by avoiding direct contact with the soft tissues. Moreover, dentists should be cautious when using sharp instruments, opting instead for blunt-tipped instruments wherever possible to prevent accidental cuts or lacerations. Special care should also be taken to maintain proper suction during procedures to prevent blood pooling, which can lead to excessive

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bleeding in hemophilic patients (19). These strategies, combined with ongoing preventive care, help ensure that routine dental procedures are as safe as possible for children with hemophilia.

Prevalence and Management of Common Dental Issues in Children with Hemophilia

Children with hemophilia face a higher risk of various dental issues due to the nature of their bleeding disorder, which complicates both routine oral care and the treatment of dental problems. The prevalence of oral diseases such as dental caries and periodontal disease tends to be higher in this population, largely because the fear of bleeding episodes discourages regular dental visits and effective oral hygiene practices. Gingival bleeding, a common issue in hemophilic children, further exacerbates oral health problems by creating a favorable environment for bacterial growth and plaque accumulation (20). As a result, these children are more likely to experience significant oral health challenges that require specialized management strategies.

Periodontal disease is among the most prevalent dental issues seen in children with hemophilia. The condition is primarily driven by poor oral hygiene and the reluctance to perform routine care due to the fear of triggering gingival bleeding. Without proper oral hygiene, plaque and tartar buildup on teeth leads to gingival inflammation, which can cause spontaneous bleeding and further complicate periodontal health. Periodontitis, the more advanced form of periodontal disease, can develop if left untreated, leading to bone loss and tooth mobility. To manage periodontal disease in hemophilic children, dentists need to focus on preventive care, including the use of soft-bristled toothbrushes and gentle flossing techniques to reduce trauma to the gums. Regular professional cleanings are also crucial, although these may require the administration of clotting factor concentrates or antifibrinolytics to control bleeding during the procedure (3, 21).

Dental caries is another significant issue among children with hemophilia, often resulting from inconsistent oral hygiene practices and frequent consumption of sugary medications. The hesitancy to brush and floss properly due to bleeding concerns can lead to increased plaque accumulation, which in turn raises the risk of tooth decay. In addition, many hemophilic children take liquid medications that are high in sugar content, further contributing to the development of dental caries. Managing caries in this population requires a balance between effective treatment and minimizing bleeding risks. Dentists should prioritize minimally invasive techniques, such as the use of fluoride treatments and sealants, to prevent the progression of caries without the need for more invasive restorative procedures (22-24).

Gingival bleeding itself is both a symptom and a cause of further oral health problems in hemophilic children. It often occurs spontaneously or following minor trauma, such as brushing or eating hard foods, and can persist for extended periods due to the patient's impaired clotting mechanism. This prolonged bleeding not only causes discomfort but also increases the likelihood of infection and complicates the maintenance of good oral hygiene. Management of gingival bleeding in hemophilic patients typically involves a combination of clotting factor replacement therapy and local hemostatic measures, such as applying pressure to the bleeding site or using hemostatic agents like tranexamic acid. Dentists may also recommend chlorhexidine mouthwash to reduce bacterial load and minimize the risk of infection in areas where bleeding is recurrent (7, 25).

The presence of oral ulcers is also common in hemophilia, which may develop due to trauma or spontaneously in areas of the oral mucosa. These ulcers can be particularly problematic, as they may bleed extensively in hemophilic patients and take longer to heal. The management of oral ulcers in hemophilia involves not only addressing the underlying cause, such as friction from braces or biting, but also providing symptomatic relief with topical anesthetics and promoting healing using mouth rinses. Careful monitoring is essential to prevent secondary infections, and in severe cases, clotting factor replacement therapy may be necessary to control bleeding from ulcerated areas (26).

http://dx.doi.org/10.52533/JOHS.2024.41107

Conclusion

Managing the dental care of children with hemophilia requires a comprehensive approach that addresses both the underlying bleeding disorder and the associated oral health challenges. Preventive strategies, careful planning of dental procedures, and collaboration between dental and medical professionals are essential to minimize bleeding risks and maintain oral health. By employing tailored treatment approaches, dentists can effectively manage common issues like gingival bleeding, periodontal disease, and dental caries. Continued advances in dental care techniques for hemophilic patients will further improve outcomes and quality of life for these children.

Disclosure

Conflict of interest

There is no conflict of interest.

Funding

No funding

Ethical Consideration

Not applicable.

Data availability

Data that support the findings of this study are embedded within the manuscript.

Author Contribution

The authors contributed to conceptualizing, data drafting, collection and final writing of the manuscript.

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