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Case Report

Hemophilia B in a 7-year-old boy undergoing deciduous tooth extraction: A case report

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Abstract

Haemophilia B (Factor IX deficiency) also known as Christmas disease is a rare X-linked recessive bleeding disorder characterized by prolonged bleeding due to deficient clotting factor IX. Dental extractions in haemophilic patients pose a significant risk of excessive bleeding and require specialized management. This report describes the case of a 7-year-old boy with severe Haemophilia B scheduled for the extraction of a lower right deciduous incisor. Preoperative and postoperative management strategies, including Factor IX replacement therapy, local haemostatic measures, and antifibrinolytics, are discussed.

Keywords: Haemophilia B, Factor IX, Christmas Disease, Antifibrinolytics

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1. Introduction

Bleeding disorders (hypocoaguable states) can be classified into platelet disorders, coagulation factor deficiencies, mixed platelet and coagulation disorders, vascular disorders and fibrinolytic defects Haemophilia which falls in the second category is clinically characterised by prolonged clotting time and excessive bleeding into the mucosa, soft tissues, muscles and weight bearing joints[1].

Haemophilia is further classified into hemophilia A,caused by a deficiency of factor VIII and is present in 80 to 90 % of cases (1:5000births), haemophilia B caused by a deficiency of factor IX present in 1:30000 births and hemophilia C (Rosenthal syndrome) which is due to deficiency in factor XI which is very rare. There is a fourth type of haemophilia proposed by the Norwegian physician Owren in 1947, the Owren's disease or parahaemophilia, caused by a deficiency of factor V which has an incidence of one case per million children[2].

2. Causes and Inheritance

Haemophilia A is caused due to the mutation in the F8 gene located on the X chromosome Xq28.The F8 gene encodes Factor VIII, a crucial protein in the blood clotting process Haemophilia B is caused by the mutation in the F9 gene located on the X chromosome (X9 27.1-27.2).The F9 gene is responsible for encoding factor IX, another essential clotting factor[1,2]

Haemophilia C is caused by the mutation of F 11 gene located on chromosome 4q35.2, which encodes Factor XI, a protein involved in blood clotting.

Owren's disease is caused by a mutation in the gene F5 located on chromosome 1q24.2 responsible for encoding Factor V which plays an important role in the coagulation cascade.

In rare cases, spontaneous mutations can occur, meaning there is no family history of hemophilia. The severity of the condition is classified based on the percentage of clotting factor present in the blood[3].

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Severe hemophilia: <1% factor activity — frequent spontaneous bleeding.

Moderate hemophilia: 1–5% factor activity — bleeding after minor injuries or surgeries. Mild hemophilia: 5–40% factor activity — bleeding typically occurs only after significant trauma or surgeries.

3. Case Presentation

A seven year old male patient accompanied by his parents came to the department of Pediatric Dentistry of our institution with a chief complain of discomfort and mobility of the lower right primary incisor for the past few days. On taking the history it was found that the patient was diagnosed with moderate Hemophilia B (Factor IX activity 1–5%) at age 3 after excessive bleeding from a minor injury. The parents also informed that there was no known history of haemophilia in both their families.

On intra oral examination it was seen that the lower right deciduous incisor was mobile (Grade III mobility) and non-carious, but extraction was required due to interference with the eruption of the permanent successor. There was no inflammation in the gingiva which was healthy.

The patient was well-nourished with no signs of acute illness.

Figure 1: Patient suffering from hemophilia B with cannula for Factor IX infusion

Figure 2: Instruments for extraction of 81

Figure 3: Extracted 81

Figure 4: Postoperative bleeding conteolled

3.1. Investigations done[4]

Complete blood count which was found to be normal
Activated partial Thromboplastin time was prolonged (55 Seconds)
Factor IX Activity Assay was 6% which was moderately reduced
Diagnosis was moderate Hemophilia B requiring specialized preoperative planning.

3.2. Preoperative Management

Factor IX replacement therapy

Recombinant Factor IX infusion (50–60 IU/kg) was given 1 hour before extraction to achieve at least 40–50% normal Factor IX levels[5,6].

A second dose may be required 12–24 hours post-extraction for sustained hemostasis.

3.3. Antifibrinolytic therapy

Tranexamic acid (15–25 mg/kg, 3 times daily) was started preoperatively and continued for 5–7 days postoperatively to prevent clot dissolution.

3.4. Local hemostatic measures prepared

Surgical hemostasis: Minimal trauma extraction technique was used.

Pressure gauze application: Post-extraction pressure for at least 10 minutes.

3.5. Topical hemostatics

Fibrin sealant or Gelatin sponge (Gelfoam) was placed in the socket.
Sutures if necessary to secure hemostasis.

3.6. Multidisciplinary coordination[7]

Pediatric hematologist was consulted.
Hemostasis monitoring was planned (Factor IX levels & a PTT monitoring post-extraction).
A cannula was inserted into the dorsal metacarpal vein for infusing recombinant Factor IX (**Figure 1**).

3.7.. Procedure (Tooth Extraction)[1]

A simple extraction was performed using a minimally traumatic technique (**Figure 2** and **Figure 3**).
Bleeding was controlled with pressure, fibrin sealant, and a haemostatic dressing[8,9].
The patient was monitored for 30–60 minutes post-procedure for signs of bleeding (**Figure 4**).

3.8. Postoperative anagement7

3.8.1. Factor IX infusion

Second dose was given 12 hours post-extraction.

3.8.2. Antifibrinolytic therapy

Tranexamic acid was continued for 5–7 days.

3.9. Oral care instructions

The patient was asked to avoid vigorous rinsing, hot food, and to use straw for 48 hours. The patient was told to eat a soft diet for a few days and maintain gentle oral hygiene to prevent infection.

4. Monitoring for Delayed Bleeding[5,7]

Parents were advised to observe for oozing or excessive bleeding. Emergency contact was provided for unexpected post-extraction bleeding.

5. Discussion

Dental extractions in haemophilic patients require specialized management due to the risk of excessive bleeding. This case emphasizes the importance of preoperative Factor IX replacement, local haemostatic measures, and antifibrinolytic therapy to ensure a safe and successful procedure.

6. Key Considerations for Dental Management in Hemophilia B Patients[5,7]

1. Avoid NSAIDs/aspirin (increase bleeding risk).
2. Preoperative Factor IX replacement is mandatory in severe cases.
3. Antifibrinolytics (Tranexamic acid) play a crucial role in preventing postoperative bleeding.
4. Minimally invasive techniques reduce trauma and bleeding risk.
5. Close post-extraction monitoring is essential to detect delayed bleeding.

6.1. New advances

Gene therapy is emerging as a potential curative option for Hemophilia B[10].
Goal: To provide a long-term or potentially curative solution by introducing a functional F9 gene into the patient's liver cells, enabling them to produce FIX naturally.

6.2. Mechanism[10]

Vector Delivery:
Adeno-associated virus (AAV) vectors are most commonly used. The AAV vector carries a functional copy of the F9 gene (often a hyperactive variant like FIX-Padua).

6.3. Administration

A single intravenous infusion delivers the vector. The vector targets hepatocytes (liver cells), integrating episomally (not into the host genome).

6.4. Expression

The cells begin producing functional FIX, reducing or eliminating the need for external infusions.

6.5. Key advances[10]

FIX-Padua variant: A modified gene that results in a FIX protein with 8–10 times higher activity.
Clinical trials have shown sustained expression and reduction in bleeding episodes.

6.6. Notable trials

AMT-060 and AMT-061 (Etranacogene dezaparvovec): Showed long-term efficacy with good safety profiles.
SPK-9001 (Fidanacogene elaparvovec): Promising results with increased FIX levels and fewer bleeds.

6.7. Advantages[10]

1. Potential single-dose lifelong therapy.
2. Eliminates need for frequent infusions.
3. Reduced bleeding episodes and improved quality of life.

6.8. Challenges[10]

1. Immune response to AAV capsid or transgene.
2. Pre-existing AAV antibodies can limit eligibility.

3. Liver toxicity (elevated transaminases).
4. Long-term durability and safety are still under observation.

6.9. Future directions

1. Enhancing vector efficiency and safety.
2. Using alternative delivery systems (e.g., non-viral vectors).

7. Conclusion

This case highlights the challenges and considerations in managing a dental extraction in a child with severe Hemophilia B. With a multidisciplinary approach, proper preoperative preparation, and postoperative monitoring, invasive dental procedures can be safely performed in hemophilic patients.

8. Source of Funding

None

9. Conflict of Interest

None.

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