

7-1-2019

## Inhibitor in hemophilia and its management-A case report

Anjalin D'Souza Ms

*Manipal College of Nursing, Manipal Academy of Higher Education (MAHE), Manipal, Udupi District, Karnataka, India, anjeline.d@manipal.edu*

Dinesh M. Nayak Dr

*TMA Pai Hospital, Melaka Manipal Medical College, Manipal Academy of Higher Education, Manipal, Karnataka, India*

Annamma Kurien Dr

*Melaka Manipal Medical College, Manipal Academy of Higher Education, Manipal, Karnataka, India*

Follow this and additional works at: <https://impressions.manipal.edu/mjnhs>



Part of the [Nursing Commons](#)

---

### Recommended Citation

D'Souza, Anjalin Ms; Nayak, Dinesh M. Dr; and Kurien, Annamma Dr (2019) "Inhibitor in hemophilia and its management-A case report," *Manipal Journal of Nursing and Health Sciences*: Vol. 5: Iss. 2, . Available at: <https://impressions.manipal.edu/mjnhs/vol5/iss2/10>

This Case Report is brought to you for free and open access by the MAHE Journals at Impressions@MAHE. It has been accepted for inclusion in Manipal Journal of Nursing and Health Sciences by an authorized editor of Impressions@MAHE. For more information, please contact [impressions@manipal.edu](mailto:impressions@manipal.edu).

# Inhibitor in hemophilia and its management-A case report

Anjalín D'Souza\*, Dinesh M Nayak, Annamma Kurien

Email: anjeline.d@manipal.edu

## Abstract

Hemophilia is a genetic disease with a deficiency of factor VIII or IX. People with inhibitors require special care and treatment due to complexity of the disease. Treatment is expensive due to high dose infusion and unavailability of factor in developing countries like India. This case report focuses on role of comprehensive care and follow-up of an adolescent with inhibitors.

**Key words:** Children, factor VIII deficiency, hemophilia, inhibitors

## Introduction

Hemophilia is an inherited X-linked coagulation disorder caused by deficiencies of the clotting factor VIII (FVIII: hemophilia A) or factor IX (FIX: hemophilia B). In India, 18,353 people are diagnosed to have hemophilia as per the recent annual global survey report (WFH, Annual Global Survey 2016, 2017). Hemophilia is classified based on percentage of factor levels as mild, moderate and severe in clinical severity. Spontaneous bleeds are more in moderate and severe hemophiliacs compared to people with mild hemophilia (Phadke, 2011).

Inhibitors are the antibodies produced by the body to fight off the foreign protein present in the treatment product. People with severe hemophilia have got more risk of developing inhibitors than mild and moderate hemophilia. Inhibitors occur among 25-30% of children

with severe hemophilia A and 1-6% of individuals with hemophilia B. Regular testing for inhibitors should be done among newly diagnosed people with hemophilia between the 1st and 50th days of treatment. Check for inhibitor at least twice a year after the 50th day of treatment, until they have received 150-200 doses. Continue checking for inhibitor once a year after that. Before any major surgery also inhibitor needs to be checked. When the person is not responding to standard treatment usually inhibitor is suspected (WFH, 2014).

## Case report

A 13-year old boy with factor VIII deficiency was admitted with swelling of the left knee joint. He was diagnosed with hemophilia at the age of two years and on-demand factor replacement therapy. Initially he was treated with skin traction, analgesics and tranexamic acid. But the symptoms did not subside with the treatment. Blood investigations were done to find out the reason for not responding factor replacement therapy. APTT value was 85.9 Sec and inhibitor screen was positive and further Bethesda assay showed 2.1 BU as low responding inhibitors. Later he was treated with high dose factor. The family belonged to lower socioeconomic status and they had another child with hemophilia. On discharge child was stable and was advised to come for physiotherapy. The nurse coordinator played a significant role in educating the

**Anjalín D'Souza<sup>1</sup>, Dinesh M Nayak<sup>2</sup>, Annamma Kurien<sup>3</sup>**

1. Assistant Professor, Manipal College of Nursing, Manipal Academy of Higher Education (MAHE), Manipal, Udupi District, Karnataka, India
2. Professor, Department of Pediatrics, TMA Pai Hospital, Melaka Manipal Medical College, Manipal Academy of Higher Education, Manipal, Karnataka, India
3. HOD, Department of Pathology, Melaka Manipal Medical College, Manipal Academy of Higher Education, Manipal, Karnataka, India

Manuscript received: 23 January 2019

Revision accepted: 09 May 2019

\*Corresponding Author

**How to cite this article:** Dsouza, A., Nayak, D. M., Kurien, A. (2019). Inhibitor in hemophilia and its management - A case report. *Manipal Journal of Nursing and Health Sciences*, 5(2). 56-58.

family members about prevention of injuries, inhibitor and its management and regular follow-up.

### Discussion

Inhibitors make the treatment difficult and complicated mainly due to financial constraints and the availability of factors. Inhibitors are diagnosed by activated partial thromboplastin time (APTT) and confirmatory tests like Bethesda assay, or Nijmegen method (Verbruggen, Van Heerde, & Laros-van Gorkom, 2009). Inhibitor levels are categorized as 'high titre' (more than 5 Bethesda Units) or 'low titre' (less than 5 Bethesda Units). 'Low responding' and 'High responding' is one of the classifications of inhibitors based on how the person's immune system reacts to factor concentrates (Giuffrida, et al., 2008).

Treatment for a person with the inhibitor is decided based on inhibitor titre and anamnestic response. We also need to consider the site and severity of the bleed. Factor concentrates can be administered at higher doses and/or more frequent intervals for acute bleeding in low responders. Acute bleeding in people with high titre inhibitors are treated with bypassing agents, such as activated prothrombin complex concentrates (APCC) and recombinant factor VIIa (Novoseven) (Franchini & Mannucci, 2011). Factor Eight Inhibitor Bypassing Agent (FEIBA®) is an example for APCC which is made from human plasma and contains variable amounts of clotting factors. This is given once in eight to twelve hours but should be limited to low doses such as 100 U/kg per infusion and no more than 200 U/kg/day. Blood clot formation is a risk which has been associated with excess use of FEIBA. High cost and unavailability of the treatment products in every country are the two main difficulties for patients with inhibitors.

rFVIIa (NovoSeven®) is a synthetic product and needs to be administered once in two to three hours. An antifibrinolytic drug such as Tranexamic acid also can be used for better outcomes. A single-centre, mixed retrospective and prospective study showed that

Immune tolerance induction therapy was successful in treating severe hemophilia A patients (Haya, et al., 2019). Regular rehabilitation program helps the individuals with inhibitors as they have higher risk for bleeding. Rehabilitation program needs to be tailored based on age, joint score and other co-morbidities (Kubota, et al., 2018).

### Conclusion

The inhibitors occur later in life in severe hemophilia A and their development is generally associated with a change in bleeding pattern. A range of therapeutic options to manage the inhibitor is currently available and comprehensive nursing care is needed to manage teenagers with inhibitors. They need to be emphasized on pain management, regular treatment and screening of inhibitors.

Source of support: None

Conflict of interest: None declared

Source of support in the form of grants: None

### Reference

- Franchini, M., & Mannucci, P. M. (2011). Inhibitors of propagation of coagulation (factors VIII, IX and XI): a review of current therapeutic practice. *72*(4), 553-62. doi:10.1111/j.1365-2125.2010.03899.x
- Giuffrida, A. C., Genesini, S., Franchini, M., Gironcoli, M. D., Aprili, G., & Gandini, G. (2008). Inhibitors in mild/moderate haemophilia A: two case reports and a literature review. *Blood Transfus*, *6*(3), 163–168.
- Haya, S., Solano, C., Cid, A. R., et al. (2019). Predictive factors of immune tolerance treatment response in severe haemophilia A patients with inhibitors: A real-world report from a single centre, mixed retrospective-prospective long-term study. *Haemophilia*, *25*(2), e97-e100. doi:10.1111/hae.13660
- Kubota, M., Takedani, H., Ono, H., et al. (2018). A case report on a multicentre cooperative rehabilitation programme for inhibitor-positive patients with haemophilia A. *Haemophilia*, *24*(4), e248-e252.

- Phadke, S. (2011). Hemophilia Care in India: A Review and Experience from a Tertiary Care Centre in Uttar Pradesh. *Indian J Hematol Blood Transfus*, 27(3), 121–126. doi:10.1007/s12288-011-0084-0
- Verbruggen, B., Van Heerde, W. L., & Laros-van Gorkom, B. A. (2009). Improvements in factor VIII inhibitor detection: From Bethesda to Nijmegen. *Semin Thromb Hemost*, 35(8), 752-9. doi:10.1055/s-0029-1245107
- WFH. (2014, December). *Who is at risk of developing inhibitors?* Retrieved May 09, 2019, from WFH Network: <https://www.wfh.org/en/page.aspx?pid=653>
- WFH. (2017, October). *Annual Global Survey 2016*. Retrieved from WFH Network: <https://www1.wfh.org/publications/files/pdf-1690.pdf>