Inhibitor in hemophilia and its management-A case report

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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