EXPERIENCE OF PROPHYLAXIS WITH ACTIVATED PROTHROMBIN COMPLEX CONCENTRATE IN A CHILD WITH HEMOPHILIA B AND INHIBITOR

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INTRODUCTION and PURPOSE

The development of inhibitor in patients with hemophilia B is a rare and severe complication with limited therapeutic options. Immune tolerance treatment is very critical option to use for hemophilia B with inhibitor. Management of the bleeding in patients with factor IX inhibitor is complicated because activated prothrombin complex concentrates (APCC) contain FIX and can cause severe allergic reactions. Recombinant activated factor VII (rVIIa) appears to be the only logical treatment for FIX inhibitor in patients with acute bleeding episodes.

METHODS

The patient was diagnosed with severe FIX deficiency at 7 months old. He was treated on demand treatment using plasma derived factor IX. Inhibitor (2,5 BU/ml) was detected at 9 year of age. Last six years, on-demand treatment was used by only recombinant faktor VIIa. In this period recurrent bleeding episodes were observed and inhibitor level increased to max 70 BU/ml. His very frequent bleeding episodes were managed using rVIIa. To approach his rVIIa unresponsive bleeding episodes and recurrent iliopsoas muscle hematomas, prophylaxis with APCC, (60 U/kg/dose three times per week) was initiated at 16 years of age. At the beginning of the therapy, desensitization protocol with APCC was administered because of the existing allergic reaction to FIX concentrate, premedication and dilution of APCC. APCC prophylaxis has been successfully using. After APCC prophylaxis, joint and muscle bleeding episodes were decreased and there was no allergic reaction or other adverse event after initiation of this prophylaxis, and inhibitor levels decreased to 10 BU/mL in 13 months duration.

CONCLUSIONS

To manage bleeding episodes in hemophilia B patient with inhibitor can be difficult. APCC may be one of the therapeutic options in treatment and prophylaxis. Allergic reaction and anaphylaxis should be kept in mind.