PROPHYLACTIC TREATMENT WITH ACTIVATED PROTHROMBIN COMPLEX CONCENTRATE IN A PATIENT WITH SEVERE FACTOR X – FX- DEFICIENCY

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BACKGROUND

Congenital FX deficiency is a rare bleeding disorder characterized by autosomal recessive inheritance. The incidence of homozygotes in general population is 1:1´000.000. Patients with FX deficiency may present with bleeding at any age, ie joint and muscle-bleeding, severe postoperative hemorrhage and perinatal intracranial hemorrhage

PATIENTS AND METHODS

We describe a female child treated at our hematology service, 39 days old, who showed intracranial hemorrhage with a measured level of X factor: 0.7%. After this episode, the infant presented 3 central nervous system bleeding. Prophylaxis started with fresh-frozen plasma (FFP) 2 times a week, but due to allergic reactions and no country X Factor concentrates availability, prophylaxis begins with aPCC 30 u / kg / day, 3 times a week, achieving a FX level of 127%. The levels of factor II, VII and IX were normal before and after administration of the aPPC. No bleeding episodes have occurred again and the patient has a normal neurodevelopmental 4 years after aPCC prophylaxis

CONCLUSIONS

Treatment options for patients with FX deficiency include FPP administration, aPCC (containing factors II, VII, IX and X) and concentrate of plasma-derived Factor X. However, aPCC can be a good alternative treatment. Currently in the literature there are few reported cases of prophylaxis in Patients with aPPC with severe FX deficiency.

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