Variable Success with Immune Suppressive Therapy in Rescuing Immune Tolerance Induction in Children with Severe Haemophilia A, FVIII inhibitors and unfavorable prognostic factors

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Epidemiology of haemophilia

- In severity structure of hemophilia A a severe form of the disorder prevails. Severe hemophilia is detected in 65% of 113 children under 18 years old suffering from hemophilia.
- Inhibitor form occurred in 14 out of 74 children with severe hemophilia A what amounts 18,9 %.
- Prevalence of hemophilia A since 1996 till 2014 is 11,55 for 100 000 boys, inhibitor form of hemophilia - 1,43.
- Increase (decrease) rate of prevalence of hemophilia without regard for severity is (-1,12%).
- Increase (decrease) rate of prevalence of inhibitor form of hemophilia (-2,3%) passed ahead the decrease of prevalence of hemophilia.

Treatment of children with inhibitor form of hemophilia A

ITI – 5 children at the age of 12 years from 2011 till 2015 with good results;
1 child at the age of 15 years – has been continuing treatment for 7 month.

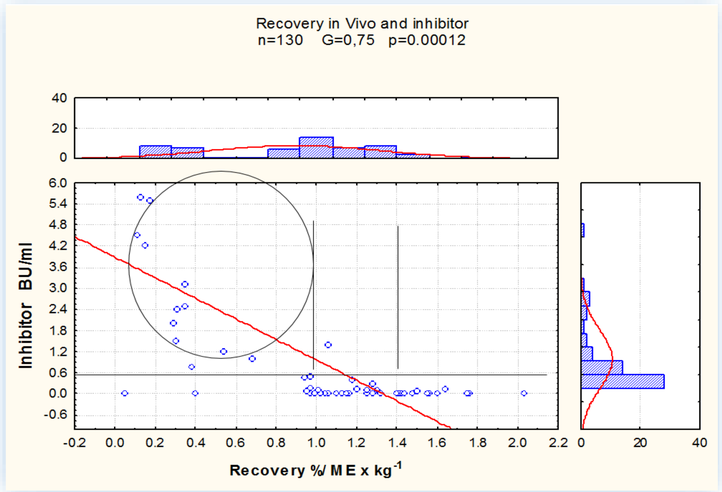
- Administration of the concentrates of prothrombin complex factors with bypass activation mechanism of blood coagulation to prevent hemorrhage at the stage of immunological tolerance induction can be initiated on actual basis of spontaneous hemorrhagic complications.
- If the residual level of factor VIII is more than 1%, the probability of a spontaneous hemorrhage is minimal, prophylactic administration of the concentrate of prothrombin complex factors with bypass activation mechanism of blood coagulation to prevent spontaneous hemorrhage is not required.

Prevention of hemorrhage at the stage of ITI

Depending on the case, 12 hours (24 or 48 hours) before the visit, a reduced dose of the product is administered to the patient, after that during the next visit the residual level and retrieval of coagulation activity are estimated 15 minutes after a repeated administration of a reduced dose. After analyzing the resulting data they evaluate the need to maintain or change the doses and frequency of factor VIII administration. The criteria of adequacy of the dose administered is the residual level of factor VIII in the patient's blood at the level of more than 1% and the recovery rate no less than 1.5-1.6% /ME kg-1.

Reducing the daily dose and increase of the administration interval

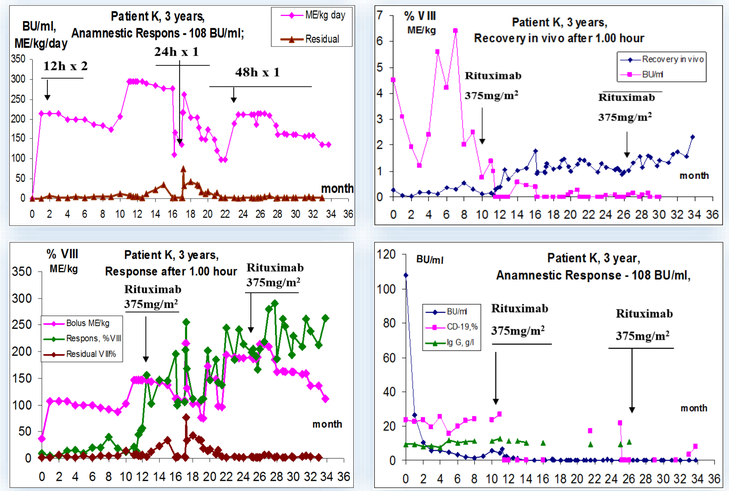
The criterion, determining the possibility of reducing the next dose of the product, or increase the interval between administrations is retrieval of coagulation activity by 70-80%, which corresponds to a recovery rate from 1,4 % / ME kg -1 or more in the absence of inhibitors.



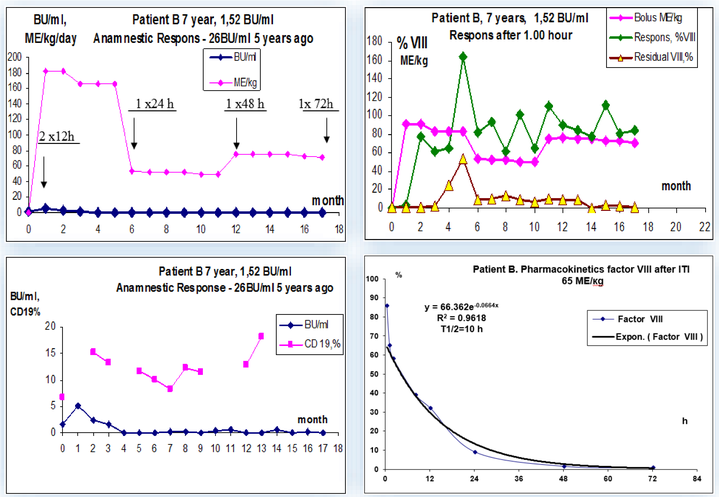
Rationale for the use of "Rituximab" medical product for ITI

Indication for the using of "Rituximab" in cases of inhibitor form of congenital hemophilia is the loss of coagulation response to the increasing administrated dose of factor VIII affected by the immune inhibitors titer increase. In some cases repeated administration of the product is allowable.

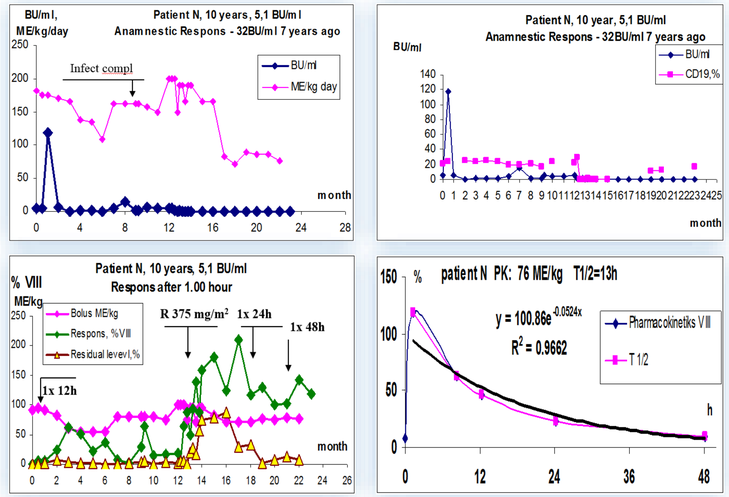
ITI in patient K



ITI in patient B



ITI in patient N



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