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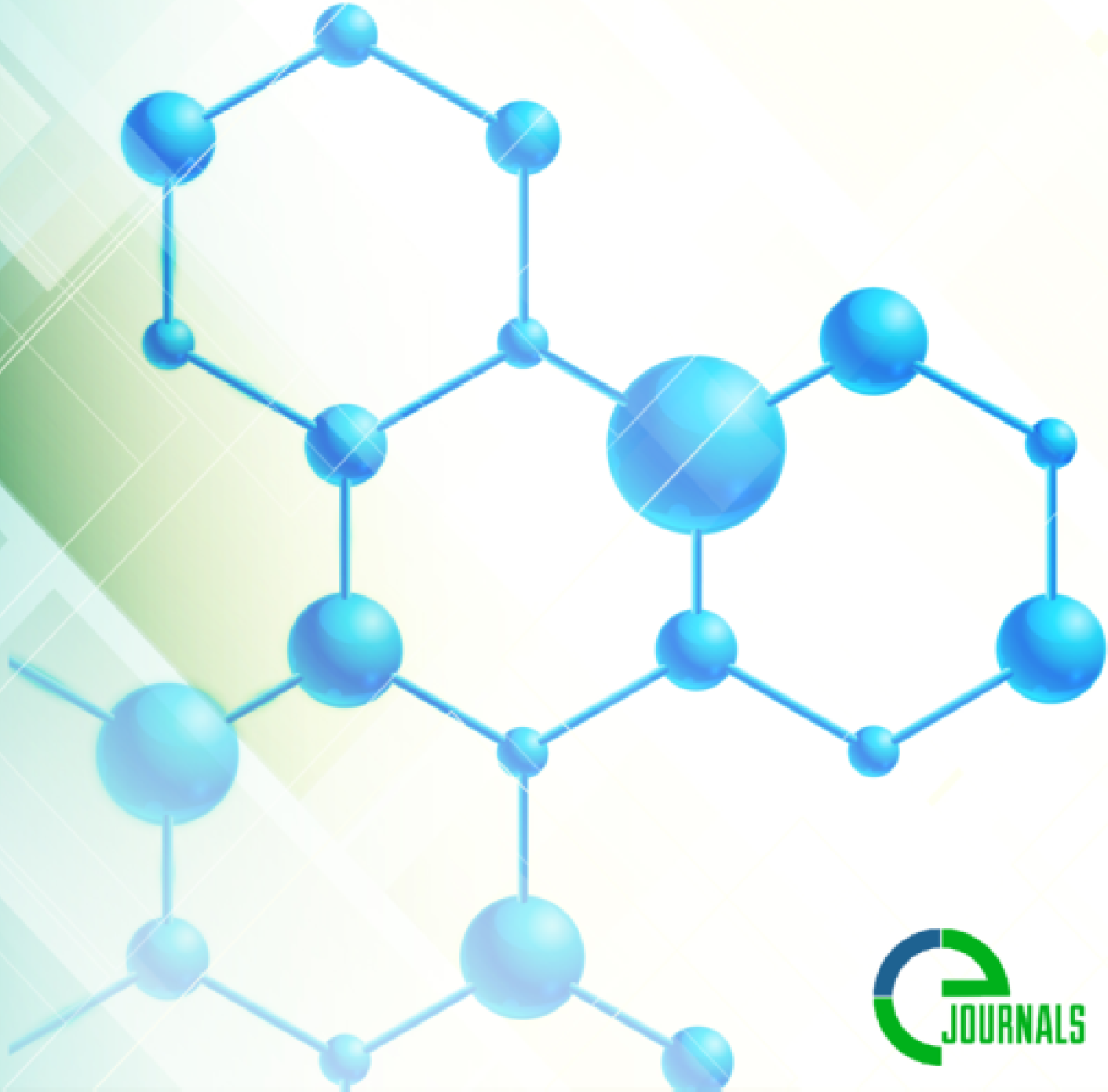

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THE IMPORTANCE OF THE LEVEL OF FACTOR VIII IN PATIENTS WITH HEMOPHILIA UNDER THE DEPENDENT THERAPY BY THE PREPARATIONS OF THE NEW GENERATION BLOOD FACTORS. REPUBLICAN SPECIALIZED SCIENTIFIC AND PRACTICAL MEDICAL CENTER OF HEMATOLOGY OF THE MINISTRY OF HEALTH OF THE REPUBLIC OF UZBEKISTAN

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Abstract: The aim of this work is to study the level of factor VIII in patients with hemophilia during replacement therapy with plasma drugs of blood coagulation factors.

In the treatment of patients with hemophilia, the main component is timely adequate lifelong replacement therapy, which makes it possible to replenish the level of the deficient factor in the plasma. The main goal of such hemostatic therapy is to increase the content of the deficient factor in the patient's blood to an effective level. The optimum hemostatic effect of factor VIII is within its concentration from 50 to 100%, and factor IX - 40-60%.

Keywords: hemarthrosis, coagulopathy, hemorrhage, blood coagulation.

Introduction.

Hemophilia is a rare hereditary disease associated with impaired coagulation (blood clotting). Scientists attribute hereditary factors to the main reasons provoking the development of hemophilia. Genetically defective blood clotting is inherited from generation to generation, and the carrier of the defective gene is exclusively the female organism, and the patient with hemophilia, as a rule, turns out to be a man. In fairness, it is worth noting that there are scientifically described cases of hemophilia in women, but these cases are extremely rare and occur when both parents of a sick girl are carriers of the damaged gene. Passing the disease on to her children, the female "conductor" herself remains healthy, her sons are doomed to hemophilia, and her daughters also become carriers of the hidden gene until they pass it on to their children. With this disease, hemorrhages occur in the joints, muscles and internal organs, both spontaneous and as a result of trauma or surgery. With hemophilia, the risk of death of the patient from hemorrhage in the brain and other vital organs increases sharply, even with minor trauma. Patients with severe hemophilia are subject to disability due to frequent hemorrhages in the joints (hemarthrosis) and muscle tissue (hematomas). Hemophilia refers to hemorrhagic diathesis caused by impaired plasma hemostasis (coagulopathy).

Treatment of patients with hemophilia is usually based on substitution therapy, i.e. on the replacement of the missing clotting factor. In the absence of proper treatment, these children become deeply disabled by school age. Modern management protocols that

allow maintaining the health of more than 90% of patients with hemophilia are based on replacement therapy with factor VIII drugs. At the same time, in order to obtain the required effect, it is necessary to achieve and maintain a certain activity of factor VIII in the blood. In the treatment of patients with hemophilia, the main component is timely adequate life-long replacement therapy, which makes it possible to replenish the level of the deficient factor in the plasma. The main goal of such hemostatic therapy is to increase the content of the deficient factor in the patient's blood to an effective level. The optimum of the hemostatic action of factor VIII is in the range of its concentration from 50 to 100%, and factor IX - 40-60%.

In some cases, stopping bleeding can be achieved with a lower concentration of factor IX and factor VIII in the circulating blood. However, hemostasis is not effective enough, a relapse of bleeding is likely. For its prevention, a patient with hemophilia requires repeated administration of antihemophilic drugs for a long time. The effectiveness of hemostatic therapy is assessed on the basis of clinical signs and coagulological studies, which, when using replacement therapy (recombinant and plasma coagulation factors), are produced before and after the administration of these drugs. For invasive and minor surgical interventions, coagulological studies are performed within a few days after the operation. This makes it possible to calculate the half-life of biological activity (T 1/2) of factor VIII or factor IX, which is of great practical importance.

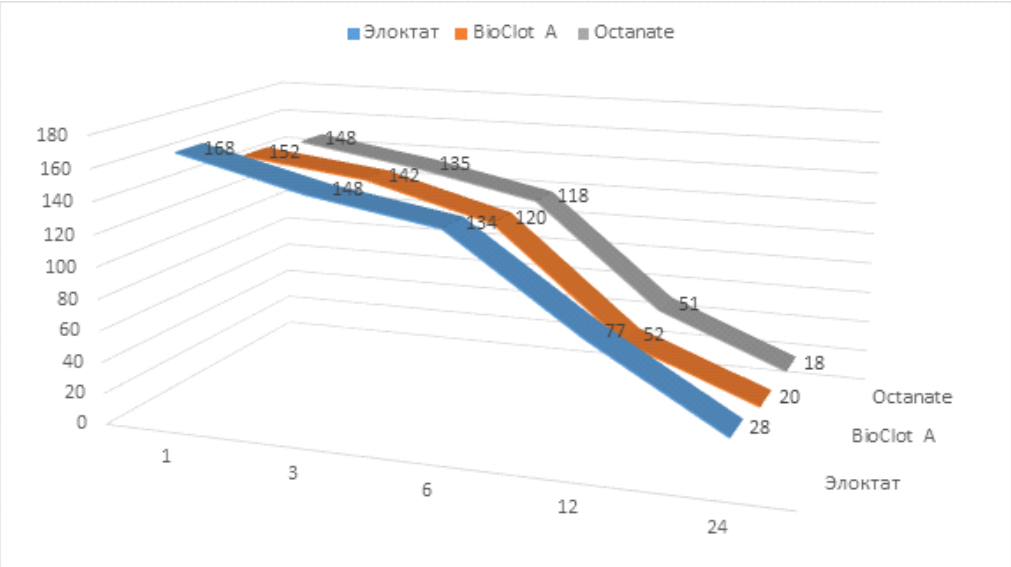
The aim of this work is to study the level of factor VIII in patients with hemophilia with replacement therapy with drugs of new generation coagulation factors.

Materials and research methods. The study is based on a clinical analysis of dynamic observations of 125 patients with hemophilia A being treated at the RSSPMCG of the Ministry of Health of the Republic of Uzbekistan. Among the examined patients - all males aged 5 to 55 years, the average age is 29.6 years. Clinical, biochemical and coagulological research methods were examined in all patients.

Results and discussion. 125 patients with hemophilia A, aged from 5 to 58 years old (median - 29.6 years) were examined. Among them, a severe course of the disease (f. VIII <1%) was found in 37 (32.7%) patients and moderate (f. VIII - 1-5%) in 76 (67.3%) patients. In order to exclude the influence on the results of measuring the level of the factor VIII administered with the preparation, the patients for some time before the study (5-8 days, on average 6.05 ± 1.02 days) did not receive other blood preparations containing factor VIII in varying amounts. The patients were divided into 3 groups. Group 1 - 51 patients aged 10 to 45 years with severe hemophilia A (factor VIII level <2.5%), to whom Eloktat was administered intravenously at the rate of 40 IU / kg of the patient's body weight. Group 2 - 36 patients with severe hemophilia A, aged 9 to 43 years, who were treated with the blood coagulation factor BioClot A. Group 3 - 38 patients with severe hemophilia who were treated with Octanate. The body weight of the patients averaged 48.65 ± 15.2 kg.

When analyzing the change in the level of factor VIII activity in the blood of patients after the administration of the calculated dose of Eloktat, individually differing curves were obtained, reflecting the dependence of the change in the factor activity on time. The maximum rise in the level of factor VIII to 168 ± 6.48% was recorded 1 hour after administration; after 3 hours the level of factor VIII decreased to 148.31 ± 4.07%, after 6 hours - to 134.29 ± 8.16%, after 12 hours - to 76.76 ± 6.89%, and after 24 hours - up to 27.58 ± 9.10% (Fig. 1)

Factor VIII activity in patients with hemophilia after administration of Eloktat, BioClot A and Octanate.



In group 2, in 36 patients who received intravenous BioClot A (40 IU / kg body weight of patients), the level of factor VIII after 1 hour rose to 152.08 ± 6.58%, after 3 hours it reached 142.34 ± 3 , 87%, after 6 hours - 119.68 ± 4.36%. After 12 hours, the factor VIII level decreased to 52.45 ± 2.46%, and after 24 hours it was 20.26 ± 1.88%.

In group 3 patients who received Octanate, the level of factor VIII after 1 hour reached 148.12 ± 6.48%, after 3 hours it was 135.26 ± 9.25%, after 6 hours it decreased to 118.18 ± 7, 95%, after 12 hours - up to 51.05 ± 6.31%, after 24 hours - up to 18.1 ± 1.13%.

The use of recombinant factor VIII - Eloktat, plasma factors BioClot A and Octanate showed a high hemostatic effect, characterized by a rapid recovery of the function of the affected limb, relief of pain, which made it possible in 97 (77.6%) patients. for the treatment of acute hemarthrosis in almost one step. In 28 (22.4%) patients, in the treatment of recurrent hemarthrosis and hematomas, only weakening of bleeding symptoms was clinically obtained, requiring repeated administration of blood coagulation factor preparations with an interval of 12 hours from the first administration within 3-4 days. There were no adverse reactions to the administration of the drug.

We also studied other indicators of hemostasis before and one hour after drug administration. The obtained results of the coagulogram are presented in table 1. Table 1.

Indicators of the coagulation link of hemostasis in patients with hemophilia A before and after the introduction of replacement therapy (M ± m)

Coagulogram indicators	Patients with hemophilia (n=125)	
	Before introduction	After injection (after 24 hours)
APPT	98,5± 9,34	39,6± 6,43*
PT, %	103,5±2,03	107, 8±1,40
TT, sec	17,2±0,32	15,4±0,28
Fibrinogen, mg / dl	6,48±0,48	4,03±0,30*
Fibrinolytic activity, min	180,5±20,58	160,88±15,48
Tolerance to heparin, min	22,5±0,32	10,4±0,21
F.VIII, %	5,3±0,84	156,13±4,56*
F.VWF, %	105,9±0,72	148,04±6,48

Note: -p <0.01; -p <0.001-reliability between the indicators before and after the use of substitution therapy.

Conclusion. Thus, hemophilia A is the most classic form of hemophilia, in the pathogenesis of which the main link is a decrease in the blood plasma factor VIII, which manifests itself as a characteristic hemorrhagic syndrome in the form of prolonged bleeding.

It has been proven that the drugs Emoclot, BioClot A and Octanate provide a therapeutic effect in the provision of highly specialized care for patients with hemophilia A with hemarthrosis of the knee joints and prolonged bleeding. It has been shown that the restoration of the activity of factor VIII in plasma after administration of these drugs occurs in accordance with the calculated parameters. Recombinant factor VIII - Eloktat and plasma factors BioClot A and Octanate are highly effective hemostatic drugs in the treatment of hemophilia A, increasing the level of the deficient factor. With a single application at a dose of 40 IU / kg of body weight, it is possible to treat acute hemarthrosis with stopping a bleeding episode, while in the treatment of recurrent hemarthrosis and hematomas, to obtain the desired result, this dose must be administered within 3-5 days.

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