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Features of the Course and Measures for the Correction of Thrombocytopathy in Adolescents

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Abstract: *This paper discusses features of the course and measures for the correction of thrombocytopathy in adolescents. Thrombocytopathy is a chronic disorder of normal hemostasis, in which the number of formed cells (platelets) remains at an acceptable level, but their functionality decreases significantly, which leads to a catastrophe on the part of the entire hematopoietic system.*

Keywords: *thrombocytopathy, syndrome, symptomatology, pathological process, mutation.*

I. INTRODUCTION

Thrombocytopathy is a chronic disorder of normal hemostasis, in which the number of formed cells (platelets) remains at an acceptable level, but their functionality decreases significantly, which leads to a catastrophe on the part of the entire hematopoietic system. The symptomatology is severe, accompanied by hemorrhagic syndrome and other phenomena, for which, in general, dangerous complications are typical. Deadly in nature 1786-1789. Treatment is required as soon as possible. In some cases, it is necessary to limit oneself to the elimination of basic symptoms, which does not completely cope with the problem, but increases the chances of survival and improves prognosis. Thrombocytopathy has an ICD-10 code - D69 with a postfix indicating the origin of the pathological process [5].

At the heart of the formation of the disorder lies a group of possible moments. In more detail: Hereditary predisposition, defects in certain genes. According to statistics, there is a considered deviation in 5-10% of cases from the total mass of the disorder on the part of the hematopoietic system and hemostasis. According to various estimates, up to 70% of clinical situations fall on the share of genetically determined thrombocytopathies. In this case, mutations are noted that exclude the possibility of a cure. It remains only to correct the symptoms. Hereditary forms are accompanied by a rapid progression of the condition, critical disorders in the functioning of all organs and systems. Decreased production of coagulation factors, adhesion and platelet aggregation. As a result of the course of hypovitaminosis, for example, a lack of B12 or B9 (megaloblastic anemia), other disorders such as DIC, current autoimmune inflammatory processes (as an option - systemic vasculitis).

In both cases, the disorder develops rapidly. However, hereditary forms of thrombocytopathy provoke the main symptoms immediately, from childhood. Due to the lack of specific signs, even experienced doctors cannot always immediately determine the essence of the pathological process. False diagnoses are made, ineffective therapy is carried out. This can cost the patient's life. As a result of violations, a complex disorder of normal hemostasis or a specific stage of it is observed. For example, adhesion (sticking), or aggregation (disaggregation thrombocytopathies). At each stage, the body involves certain substances. Violations of synthesis and is provoked by genetic mutations transferred by diseases. It is not possible to eliminate the etiological factor in all cases. Acquired forms have a clear and understandable symptomatology [2.p.42]. The development of the clinical picture can be easily linked to previous processes. Unfortunately, even when suspicious symptoms are detected, patients are in no hurry to contact specialists, lose precious time and miss the opportunity for a complete cure. Classification The subdivision is carried out for several reasons. The first and already mentioned is the origin of the pathological process.

According to him, the following types of violations are called:

- 1) Congenital form. It occurs in the vast majority of recorded and described clinical situations. Accompanied by critical disorders of hemostasis, therapy does not give a pronounced effect. For this reason, it remains for doctors to deal with the symptoms, to eliminate the most dangerous signs. Regular therapeutic measures are required. Courses 2-4 times a year. Forecasts remain vague.

- 2) Acquired type. Occurs in about 30% of situations. Restorative measures are possible, with timely detection, it is possible to completely cope with the pathological process. The main category of patients are young people from 20 to 35 years old. In the fairer sex, thrombocytopathy is found somewhat less frequently (about 1.5-2 times) [1]. Congenital form of the pathological process Assumes division depending on the stage at which hemostasis is disturbed. Problems in terms of adhesion (adhesion) of formed blood cells. Disorder of the first stage of restoring the integrity of tissues in the destruction of blood vessels. Accompanied by a decrease in the synthesis of certain substances. The most severe symptoms are characteristic, since coagulation is impossible even to a minimal extent (von Willebrand disease, macrocytic platelet degeneration of Bernard-Soulier). Violations of normal aggregation. Accompanied by insufficient production of protein structures.
- 3) Autosomal recessive or dominant disaggregation thrombocytopathy
- 4) Glyantsman's thrombasthenia. Partial disaggregation thrombocytopathies. Caused by the loss of part of the function of platelets.
- 5) Collagen aggregation, ADP, thrombin aggregation.

These include: anomaly of May-Hegglin, Pearson-Stob, hereditary afibrinogenemia. Gray platelet syndrome (with insufficient storage of granules and their components): Herzhmansky-Pudlak syndrome and other deviations of a similar plan. Separately, it is worth mentioning the pathology associated with congenital anatomical defects, genetic abnormalities. For example, heart defects, Down syndrome and other similar phenomena.

Acquired thrombocytopathies Classified according to the nature of the disorder. Accordingly, two more forms of violation are called: Acute. Episodic, rapid course of the disease, in general, is atypical. Therefore, it is relatively rare to talk about it. Violation is accompanied by a rapid unfolding of the full clinical picture, critical manifestations, the condition worsens in a matter of hours, ends in death especially often. Chronic. The most common type. Talking about the acute form of thrombocytopathy is not entirely true. Because the process is in a latent state up to a certain point, critical violations are accompanied by the onset of dysfunction, its manifestation. Symptoms The clinical picture is determined by the development of typical disorders. They correspond to violations of normal coagulability. Frequent nosebleeds. Regardless of the influence of a third-party factor. At the same time, blood pressure remains normal or below it, there are also no mechanical factors of influence, trauma. The intensity of the process is so great that in some cases one cannot do without medical help right away. It will require tamponade, the use of drugs to restore the normal state. Read about other causes of nosebleeds (frequent and episodic) in adults here, in children - here. Bruises, hematomas on the body are a mandatory, but not a characteristic sign of thrombocytopathy. Arranged in random order. This is the result of a violation of the anatomical integrity of the vessels, small capillaries (hemorrhagic syndrome). Sizes can be significant: up to several centimeters and even more. In the course of progression, the manifestation becomes more pronounced, accompanied by a gradual increase in the number of bruises. At the same time, there is no pain at the site of hematoma formation, because there was no mechanical factor - tissue damage [4.p.121].

Weakness. Violation of normal working capacity. A person tends to lie down, there is no strength to perform daily work. Muscular asthenia is present. The muscles become sluggish, it is impossible to move even for short distances. Drowsiness. During the daytime, at night. Constantly. Regardless of the amount of rest. This is a typical manifestation of iron deficiency anemia, which develops in almost all patients with thrombocytopathy, even in the early stages. The secondary pathological process progresses rapidly, in parallel with the formation of the main disorder. Tachycardia. Increase in heart rate. How pronounced - depends on the degree of developed anemia, hemoglobin deficiency. Cardiac structures do not receive enough oxygen, useful substances, they cannot work in the same rhythm. Intensification occurs as an adaptive mechanism. But it doesn't give any result.

Drop in blood pressure. 10-30 mm Hg less than the individual functional norm for a particular person. Dyspnea. Even without any physical activity. Accompanied by the inability to work, move, climb stairs. The person is in a state of forced rest. Sweating. Hyperhidrosis. Also the result of anemia. Frequent fainting. Insufficient nutrition and oxygen supply affects, including the brain. Syncopal episodes become more frequent as the underlying diagnosis progresses. Menorrhagia. In girls and adult women. Abundant menstrual bleeding, as well as an outpouring of liquid connective tissue, is possible out of connection with the cycle.

This is a deadly symptom that cannot but affect the general condition, aggravates the course of anemia, and provokes deadly complications in patients. Immediate hospitalization in a gynecological hospital is required. Massive bleeding. Gastrointestinal, pulmonary, others. They pose a critical threat to the life of the patient. They begin spontaneously, without visible provocateurs. The impossibility of normal surgical interventions due to the general fragility of the vessels. Inability to adequate spontaneous hemostasis. This is a big problem, since even dental treatment becomes an impossible task. Objectively, occult blood is found in the stool tests, what can we say about constant minor bleeding. Hematuria - traces of red blood cells in the urine, which should not be. The symptoms are extremely dangerous. The disorder cannot be ignored, otherwise thrombocytopathy will quickly provoke fatal complications.

The reasons There are only two development factors. They have already been mentioned. Genetic anomalies. They occur 3-4 times more often when compared with other factors provocateurs. The inheritance of the pathological process occurs directly or as an additional disorder with rare mutations. There is a wide group of specific syndromes that provoke certain dysfunctions. Deviations in the synthesis of coagulation factors, proteins. Some diseases that cause manifestations of thrombocytopathy to life. When there are enough formed cells, but they are not able to perform their functions - to ensure normal coagulation. These include processes such as hemorrhagic vasculitis, disseminated intravascular coagulation syndrome (DIC), megaloblast anemia due to insufficient amounts of vitamin B12 and / or folic acid, cirrhosis of the liver, and renal failure. The reasons are evaluated first. If a hereditary form of pathology is detected, a complete recovery is not necessary. Only symptomatic correction is possible. Diagnostics The examination is carried out under the supervision of a hematologist. In relatively mild cases, it is possible on an outpatient basis. But more often, hospitalization in a specialized hospital for a short time is required. Indicative list of activities: Oral questioning of the patient. Fixing all complaints. Based on the clinical picture, a number of hypotheses are put forward. You need to exclude them one by one, as the doctors progress, they discover a specific diagnosis. Collection of anamnesis. Thrombocytopathy in children is difficult to diagnose, since, as such, it is not possible to detect probable causes. The facts of a difficult course of pregnancy, family history are revealed. In older patients, a group of diseases transferred during the life period, current pathological processes is subject to assessment. These factors are taken into account when identifying the origin of the violation. General blood test (KLA). A normal platelet count is found. Often there are no specific deviations at all. Coagulogram. Informative laboratory technique. Aimed at detecting the rate of platelet clotting. With the disease under consideration, it increases significantly. Study of the morphological functions of platelets. Plays a major role in the diagnosis. Purposeful searches give good results and immediately make it possible to state the presence of a pathological process [1].

When examining children, it is recommended to go through three cycles during the entire acute phase. Even before the start of treatment, then in the process of therapy and already at its final stage. The list of events is not exhaustive, these are the most common. According to the indications, patients are referred to a gynecologist, ENT doctor, and other specialists. Depending on the prevailing symptoms. Treatment Therapy is carried out on an urgent or planned basis. In patients with hereditary forms of thrombocytopathy, complete recovery is impossible. With regard to acquired varieties, the elimination of the main process is required. As a rule, in both situations, the treatment is symptomatic, aimed at stopping the manifestations of the deviation. Several pharmaceutical groups are used. Angioprotectors. As the name implies, the task of these medicines is to strengthen blood vessels, normalize their elasticity. Such names as Anavenol, Askorutin, ascorbic acid are used. Complex, for a long time. The duration of the course depends on the doctor's prescription, the severity of the disorder. Products based on aminocaproic acid. Helps to stop bleeding. Can be applied locally. For example, with the destruction of the vessels of the nose, open wounds. Hemostatic agents as needed. Vikasol and similar. Vitamin and mineral complexes. metabolic drugs. Potassium orotate and the like. If necessary, iron or B12, folic acid preparations are prescribed. If correction of anemia is required. This allows you to remove some of the severe symptoms of the pathological process.

Diet changes play a big role. The menu should include a large number of products of plant origin, animal protein, peanuts. If necessary, a transfusion of erythrocyte mass, plasma is carried out. This is an extreme measure and does not always give a sufficient effect. The planned course of treatment is carried out 2-4 times a year, depending on the severity of the pathological process. Disaggregation thrombocytopathy requires more frequent interventions. Forecast In most cases, with adequate treatment, timely therapy, the prospects for recovery are quite favorable. Survival according to various estimates ranges from 60 to 85% or even more. Some forms of the pathological process cannot be corrected in principle. With the development of massive bleeding, a tendency to such, the chances of saving life are not so high. About 10-15%. The key to a successful cure is early diagnosis, it should not be delayed.

Especially if there are suspicious manifestations. Possible Complications {banner_banstat9} The consequences of thrombocytopathy are critically dangerous. Among the main ones: Massive bleeding. Uterine, gastrointestinal. Pulmonary and others. Accompanied by the release of a large amount of liquid connective tissue. asthenic phenomena. Up to the inability to perform daily activities at home, at work. Significantly reduce the quality of human life. Although they do not carry a mortal danger. Recovery allows you to partially stop the manifestations of the disorder. Possible severe disability and death from blood loss. Thrombocytopathy is a qualitative defect in platelets, a complex process that requires urgent treatment. Efficiency depends not only on the timeliness of the prescribed course, but also on the form of the disorder. Congenital, hereditary disorders have a genetic basis, therefore, only the removal of symptoms is possible, the prevention of fatal complications. The issue is resolved under the supervision of a hematologist.



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