

CHANGES IN BIOCHEMICAL PARAMETERS IN PATIENTS WITH HEMOPHILIA WITH MUSCLE HEMATOMAS BEFORE AND AFTER TREATMENT

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Annotation: This article we discussed about biochemical changes in blood hemophilia A is a worldwide disorder of the coagulation system. It is a male disease, but women with hemophilia are less common in communities with high levels of kinship. There are steps to achieve an optimum life for patients with hemophilia in developing countries, and awareness of the pattern of death in patients with hemophilia is a prerequisite for any health-care program.

Owing to the lack of any data on the pattern of death in patients with hemophilia from developing countries, the current study was done to address common causes of death, and the spectrum of causes of death among individuals with hemophilia A and B.

Keywords: hemophilia A, troponins ,creatine phosphokinase, aspartate aminotransferase (AST) , myoglobin, muscle hematoma.

Hemorrhagic symptoms are usually correlated with plasma factor VIII levels and range from a variety of hemorrhagic features, from spontaneous bleeding leading to death in the brain to skin ecchymosis. In coagulation studies, it is necessary to distinguish two types of hemophilia A and B and classify the severity of the disease. It is estimated that the number of undiagnosed patients with hemophilia A is significant in developing countries due to the limited means of diagnostic hemostasis and the lack of specialists in this field. Sometimes we come across cases with hemorrhagic symptoms that have not been identified by therapists. The purpose of this study was to interpret clinical and diagnostic parameters, traps, and coagulation analysis in hemophilia A. Aspartate aminotransferase (AST) is an enzyme found in all cells of the body, but mainly in the heart and liver and to a lesser extent in the kidneys and muscles. When the liver or muscles are damaged, AST is released and the AST content in the blood rises. In this regard, the activity of this enzyme is an indicator of liver damage. The analysis for AST is part of the so-called liver

test tests that diagnose abnormalities in the liver. A number of diseases lead to damage to liver cells, which increases the activity of AST. AST does not always reflect only liver damage; the activity of this enzyme can also increase in diseases of other organs, in particular, in myocardial infarction. To detect liver damage. As a rule, an AST test is prescribed in conjunction with an alanine aminotransferase (ALT) test or as part of a general liver function test. AST and ALT are considered the two most important indicators of liver damage, although ALT is more specific than AST. Blood AST is often compared with other tests, such as alkaline phosphatase (ALP), total protein, and bilirubin, to determine a specific form of liver disease. To monitor the effectiveness of the treatment of liver diseases. To monitor the health of patients taking medications potentially toxic to the liver. If AST activity increases, the patient may be switched to other medications.

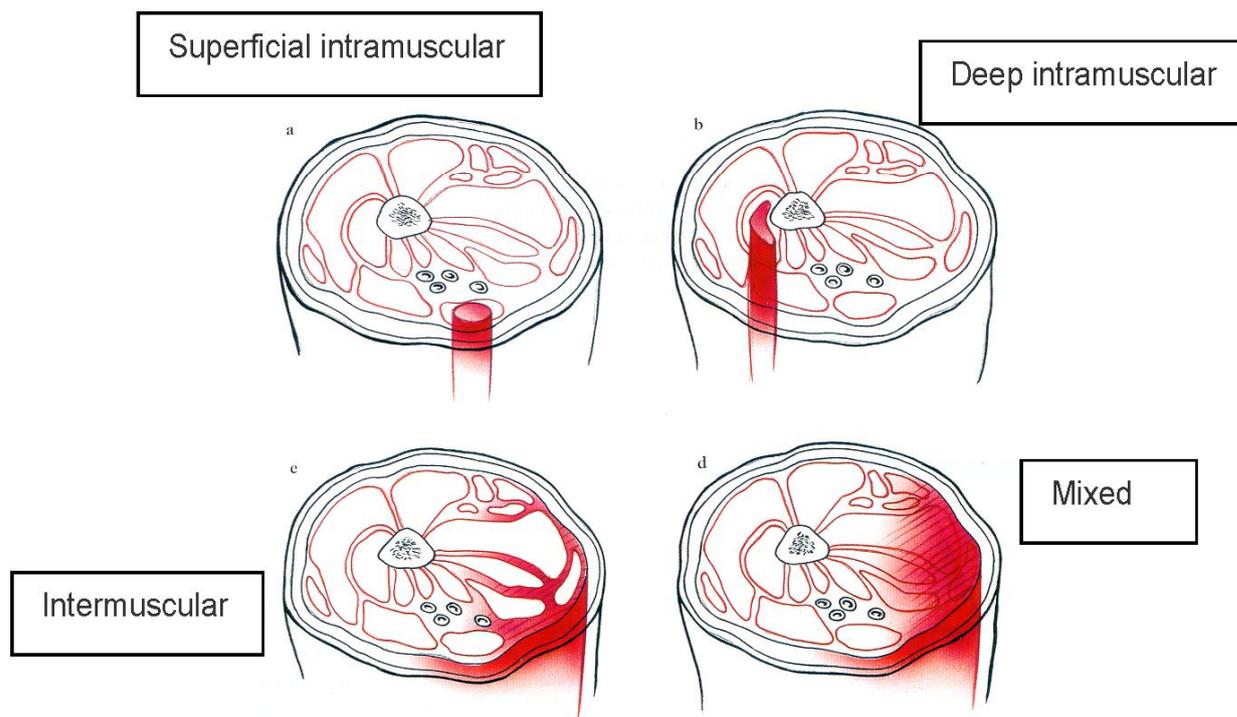
Hemophilia is a rare disorder in which your blood doesn't clot normally because it lacks sufficient blood-clotting proteins (clotting factors). If you have hemophilia, you may bleed for a longer time after an injury than you would if your blood clotted normally. Small cuts usually aren't much of a problem

Troponins are small proteins involved in the regulation of muscle contraction. Two types of troponins, troponin-I and troponin-T, are structurally different in skeletal and cardiac muscles; therefore, cardiospecific forms of troponin-I and troponin-T can be isolated by immunoassay methods. For troponins, the ratio of concentration within muscle cells to plasma concentration is much higher than for enzymes and myoglobin, which makes these proteins highly sensitive markers of myocardial damage.

Muscular hematomas are in simple terms defined by bleeding within a muscle group. These muscular hematomas may be traumatic or spontaneous. Traumatic muscle hematomas, while painful to the patient, are manageable with conservative rest and non-narcotic pain medication. Spontaneous muscle hematomas, in contrast, are mainly located in the abdominal waist area and have the potential to develop into life-threatening conditions..

Traumatic hematomas are usually treated conservatively, especially in patients who are not currently being treated with anticoagulants. In these patients, the bleeding usually stops spontaneously. In contrast, spontaneous muscular hematomas are often areas of active bleeding, and a CT angiogram may be required to better assess the hematoma and its source, as well as which vascular structure it originates from.

If possible, stopping anticoagulation is the first step in treatment and may be sufficient to ensure hemostasis and resolve muscle hematoma. Surgical evacuation of the hematoma is necessary when the neurological structures are compressed or the hematoma causes local ischemia. It should be noted that recurrences of these hematomas are common and careful monitoring is important to determine recurrence.



Intramuscular hematoma is characterized by the integrity of the epimysium and extravasation of blood into the muscular body affected by trauma. This leads to an increase in intramuscular pressure with compression of the bed of the capillaries opposite the blood vessels; therefore, clinical signs and symptoms are localized. Since the presence of blood flow can lead to an increase in the osmotic gradient, edema can increase more than 48 hours after injury. This change in the osmotic gradient forces the passage of extracellular fluid through the muscle fascia to balance the same osmotic gradient. This condition causes the injured muscle to swell in order to expand the muscle fascia or expand the boundaries of the muscle itself. The main symptoms associated with the occurrence of intramuscular hematoma are pain, especially in the first 72 hours and several days after injury, decreased contractility, as well as muscle function and expansion. The prognosis for intramuscular hematomas is worse than for intramuscular hematomas, and experts suggest treating them with drainage to avoid potential myositis ossification or fibrosis after injury. Although intramuscular hematomas initially appear more dramatic due to the resulting bruising and swelling, intramuscular hematomas are considered more serious because intact fascia increases muscle pressure.

Arterial embolization is sometimes proposed as a treatment for spontaneous muscle bleeding and has the advantage of being less invasive than surgical evacuation while maintaining a clinical success rate of 57% to 69%. About 5% of troponin-I inside muscle cells is in free form in the cytoplasm, which explains its appearance in blood plasma within 3-6 hours after damage to the heart muscle (this is also facilitated by the small size of troponin molecules). The main amount of troponin-I in the cell is associated with muscle filaments and is released slowly when the heart cell is damaged, as a result of which the increased concentration of troponin in the blood persists for 1-2 weeks after myocardial injury. The period of increased release of troponin-I thus overlaps the diagnostic windows of both creatine kinase-MB and LDH. The peak concentration of troponin-I is observed at 14-20 hours after the onset of chest pain, 7 hours after the development of acute myocardial infarction, the concentration of troponin-I is increased in 95% of patients.

After successful thrombolysis, there is a greater rise in troponin-I levels compared to patients with persistent occlusion (washout phenomenon).

The study of troponin-I is advisable when examining patients both early and late after the onset of clinical symptoms. This test is useful in deciding the choice of tactics in the management of patients with acute coronary syndrome, including patients with unstable angina. In acute coronary syndrome, an increased level of troponin-I is regarded as a sign of myocardial ischemia, caused by the activation and aggregation of platelets and leading to necrosis. An increase in troponin-I concentration in patients with unstable angina pectoris indicates a poor prognosis and the risk of developing myocardial infarction in the next 4-6 weeks. Determination of troponin-I can be used to diagnose myocardial infarction in patients with concomitant skeletal muscle injury (it has been shown that acute and chronic damage to skeletal muscles, excessive physical exertion, surgery, excluding heart surgery, muscle injuries do not cause an increase in troponin-I).

The small elevation of cardiac troponin-I must be interpreted with caution. Various pathological conditions resulting in damage to myocardial cells can potentially lead to an increase in the level of cardiospecific troponin-I. An increased level of troponin in isolation cannot serve as a basis for the diagnosis of myocardial infarction. In rare cases, troponin-I levels may increase in renal failure.

Evaluation of patients with myocardial infarction and unstable angina pectoris for prognostic purposes. Choice of treatment tactics for patients with acute coronary syndrome. Controlling the effect of chemotherapy on the myocardium.

The results of troponin-I determination should be used in conjunction with data from clinical observations and studies of other markers of myocardial damage. Serial studies of troponin-I are recommended to identify growth and decrease in its level characteristic of myocardial infarction. For dynamic monitoring of troponin-I levels during the first and subsequent blood draws, the same detection method and tube type must be used. Reference values: <1.0 ng / ml. In most cases, troponin-I is not detected in the blood. In healthy people with a positive reaction to troponin in 98% of cases, its level is below 1 ng / ml. Myocardial injury after percutaneous transluminal coronary angiography, defibrillation and other cardiac manipulations; recent unstable angina (slight rise in concentration) non-ischemic dilated cardiomyopathy; drug intoxication (cytostatics); myocarditis; rejection of a heart transplant; sepsis, and other critical (shock) conditions; end stage renal failure; duchenne-becker myodystrophy; dic syndrome. diagnosis of anemia: transferrin, ferritin, total serum iron-binding capacity (tIBC)

Anemias can be separate diseases or manifestations of some other disease. The body experiences oxygen deprivation, characterized by the following symptoms of anemia: weakness, dizziness, fainting, noise or ringing in the ears, stranded dots in the eyes (oxygen starvation of the brain); heart palpitations (the heart is forced to faster "drive" blood to compensate. lack of oxygen); shortness of breath (rapid breathing is also an attempt to compensate for oxygen starvation); pallor of the skin (especially noticeable if you pull the lower eyelid or look at the fingertips - "pale nails").

In a biochemical blood test, important indicators for the doctor and patient are:

- serum iron analysis
- iron binding capacity of blood serum (YSS)
- transferrin

- ferritin test.

The set of indicators of these analyzes most fully reflects in the diagnosis of anemia, its presence or absence, the nature of anemia.

The following isoenzymes (fractions) of creatine kinase are of greatest clinical importance: CC-MB (cardiac isoenzyme that changes when myocardial cells are damaged), CC-BB (brain isoenzyme that reflects the pathology of brain cells), CC-MM (muscle isoenzyme located in skeletal muscles).

An increase in total creatine kinase activity is observed when any of the above cells are damaged and therefore is not specific. Most often, a significant increase in the activity of this enzyme is observed in acute myocardial infarction (determination of creatine kinase, and especially the CF fraction, is widely used for early diagnosis of myocardial infarction, since its increase is noted already 2-4 hours after an acute pain attack; return of the indicator to normal occurs quickly enough (for 3-6 days), therefore, the determination of total creatine kinase in the blood at a later date for the diagnosis of myocardial infarction is not very informative). An increase in the activity of creatine kinase is often observed in acute myocarditis, however, it is not so pronounced and lasts much longer than with a heart attack.

High activity of total creatine kinase is often found in traumatic injuries and diseases of skeletal muscles (for example, with progressive muscular dystrophy, myopathy, dermatomyositis), as well as in some diseases of the brain, after surgery, taking large doses of psychotropic drugs and alcohol, with any type of shock, hypothyroidism. A decrease in the level of creatine kinase is often detected with thyrotoxicosis (increased release of thyroid hormones). Until the hematoma is completely gone, immediate (early) on-demand enhanced hematology treatment should be initiated. If left untreated, muscle bleeding can lead to complications such as nerve damage, ear syndrome, myositis ossificans, pseudotumor, and even infection (abscess). In the current literature on muscle hematomas in the non-hemophilic population, surgical drainage as an open surgical procedure may be useful in relieving symptoms better and faster if subcutaneous drainage or subcutaneous drainage fails under ultrasound control. Ultrasound hematoma evacuation is a well-tolerated procedure. However, the rate of unsuccessful evacuation and recurrence of hematoma is significant (13%). Unsuccessful evacuation rate is due to excessive density and / or stickiness of the contents. Ideally, evacuation of the hematoma should be performed 3-5 days before muscle bleeding.

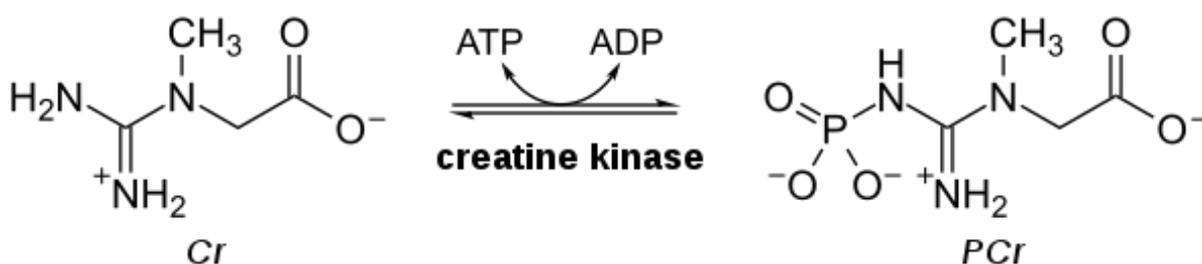
Myoglobin is an iron-containing protein found in skeletal muscle cells and in the myocardium, which supplies them with oxygen, which gives them energy to contract.

During normal functioning of the body, myoglobin in the blood is so small that it cannot be determined by laboratory methods. An increase in its concentration in the blood occurs when skeletal muscles and myocardium (heart muscle) are damaged. An increase in the level of myoglobin within 2-3 hours after the onset of pain is observed in 85% of patients with acute myocardial infarction and persists for 2-3 days (with a complicated course of a heart attack it lasts longer). It is important that the repeated rise in the level of myoglobin in the patient's blood after the normalization of the indicator reliably indicates the expansion of the infarction zone and the relapse of the disease. In addition to acute myocardial infarction, a significant increase in myoglobin in the blood is observed with extensive muscle injuries, prolonged pressure syndrome, and severe electric shock. The myoglobin molecule is formed by a single polypeptide chain and an iron-containing heme; it is similar in structure and function to blood hemoglobin.

Myoglobin functions myoglobin binds oxygen (oxymyoglobin is formed) and is the main supplier for skeletal muscle. During hypoxia (for example, during intense physical activity) oxygen is released from the complex with myoglobin and enters the mitochondria of myocytes, where ATP is synthesized. Myoglobin is excreted unchanged in the urine, so its concentration also depends on kidney function. With any damage, necrosis, lysis of skeletal muscle tissue or myocardium, myoglobin enters the bloodstream. In myocardial infarction, the severity of hypermyoglobinemia is in direct proportion to the size of the necrosis focus. This is one of the earliest markers of myocardial infarction (detected as early as 2 hours after an attack, an increase in concentration can be 10-fold), it is assumed that the rapid entry into the blood is associated with the relatively small size of the molecule, this also explains the rapid excretion of it by the kidneys from the blood

Myoglobin is a non-specific marker of myocardial infarction, therefore the diagnosis should be confirmed by more specific markers for myocardial damage. Indications: early diagnosis, monitoring and prognosis of myocardial infarction; diseases of skeletal muscles (necrosis, trauma, ischemia).

Preparation: It is recommended to donate blood in the morning, from 8 to 11 am. Blood is taken on an empty stomach, after 8-14 hours of fasting. Drinking water without gas and sugar is allowed. Food overload should be avoided on the eve of testing. Reference values: children under 18 and men: 17.4-105.7 ng / ml women: 14.3-65.8 ng / ml myocardial infarction (an increase in myoglobin level is transient, observed within 1–4 hours from the onset of symptoms, normalizes within 24 hours from the onset of an attack of pain in the heart); skeletal muscle injury; convulsions; myositis; myodystrophy; rhabdomyolysis; acute renal failure; exercise stress; burns. Decreasing the level the presence of circulating antibodies to myoglobin (with poliomyelitis); rheumatoid arthritis.



Creatine kinase (creatine phosphokinase) is an enzyme that catalyzes the formation of a high-energy compound creatine phosphate from ATP and creatine, which is consumed by the body during increased physical exertion. Contained in the cells of the heart muscle, skeletal muscles, brain, thyroid gland, lungs Such The treatment is called primary prevention. It has also been shown to be initiated after therapy the development of recurrent hemarthrosis, secondary prevention, helps to reduce severity of arthropathy. Indications for replacement therapy severe hemophilia, moderate hemophilia (factor activity less than 3%) with at least one episode of hemarthrosis or severe hemorrhagic development manifestation of other localization. Patients with factor activity greater than 3% are permanent or In case of recurrence of bleeding in the joints, long-term prophylactic treatment is necessary, symptoms of sinusitis or arthropathy, the appearance of obvious hemorrhagic manifestations, requires frequent injections of coagulation factor concentrates.

Conclusion:

Hemophilia is a worldwide bleeding disorder. It most often affects men and is rare in women. Manifestations of hemorrhage range from fatal episodes of central nervous system hemorrhage to superficial ecchymosis. Creatine kinase supplies a large amount of energy in short intervals, for example, by providing energy for muscle contractions. CPK activity is inhibited by thyroxine. In childhood, the activity of creatine kinase is higher than in adults, which is associated with the intensive growth and participation in this process of tissues rich in this enzyme - muscle and nervous. In women, the CK activity is slightly lower than in men. When cells are damaged, CK is released and enters the bloodstream. Creatine kinase is an enzyme that catalyzes the reaction of transferring a phosphoryl residue from ATP to creatine with the formation of creatine phosphate and ADP. ATP (adenosine triphosphate) is a molecule that is a source of energy in the biochemical reactions of the human body. The reaction catalyzed by creatine kinase provides energy for muscle contractions. Distinguish between creatine kinase contained in mitochondria and cell cytoplasm. The creatine kinase molecule consists of two parts, which can be represented by one of two subunits: M, from the English muscle - "muscle", and B, brain - "brain". Thus, in the human body, creatine kinase exists in the form of three isomers: MM, MV, BB. The MM isomer is contained in the skeletal muscles and myocardium, MV - mainly in the myocardium, BB - in the brain tissues, in small amounts in any cells of the body. The best way to treat hemophilia is to replace the missing blood clotting factor so that the blood can clot properly. This is typically done by injecting treatment products, called clotting factor concentrates, into a person's vein. Clinicians typically prescribe treatment products for episodic care or prophylactic care. Episodic care is used to stop a patient's bleeding episodes; prophylactic care is used to prevent bleeding episodes from occurring. Today, it's possible for people with hemophilia, and their families, to learn how to give their own clotting factor treatment products at home. Giving factor treatment products at home means that bleeds can be treated quicker, resulting in less serious bleeding and fewer side effects.

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