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# ETIOPATHOGENESIS OF HEMOLYTIC ANEMIA

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**Abstract:** Hemolytic anemias occur due to the predominance of erythrocyte destruction over formation. In hemolytic anemia, the life span of erythrocytes is reduced to 12-14 days and pathological hemolysis occurs.

**Key words:** hemolysis, erythrocyte, membranopathies, enzymopathies, hemoglobinopathies, hemolytic jaundice.

Usually, hereditary and acquired forms of hemolytic anemia are distinguished. Under physiological conditions, erythrocytes live 100-120 days. Then, the pigment free bilirubin formed as a result of the breakdown of erythrocytes by macrophages in the spleen, which loses its deformability, circulates in the blood and goes to the liver, where it binds with glucuronic acid with the help of enzymes. Bilirubin glucuronide enters the intestine as part of bile.

According to the mechanism of pathological hemolysis, it is divided into cellular and intravascular type. Intracellular hemolysis of erythrocytes takes place in the cells of the reticuloendothelial system in the spleen, and there is an increase in the amount of free bilirubin in the blood serum, an increase in the excretion of urobilinogen through urine and feces, and a tendency to the formation of stones in the gallbladder and bile ducts.

Intravascular hemolysis of erythrocytes occurs in the blood vessels with the participation of complement, hemoglobin in the plasma increases and is excreted unchanged in the urine or in the form of hemosiderin. In some cases, hemosiderin accumulates in internal organs and causes hemosiderosis. Haptoglobin formed in plasma can cause acute kidney failure by getting stuck in kidney tubules.

Hereditary hemolytic anemias are divided into 3 groups according to the location of the mutational defect:

1. Membranepathies - the erythrocyte membrane is associated with a structural disorder of protein and lipid components.
2. Fermentopathies - associated with deficiency of enzymes of erythrocyte pentose phosphate cycle, glycolysis ATF and porphyrin synthesis, nucleotides and glutathione metabolism.
3. Hemoglobinopathies - hemoglobin chain synthesis disorder.

Hereditary membranopathies include hereditary microspherocytosis - Minkowski - Shofar disease. This disease is inherited in an autosomal dominant manner. In Minkowski-Shofar disease , the membrane defect consists in the high permeability of the erythrocyte shell to $Na^+$ ions. When the $K^+$ - $Na^+$ pump is activated, the intracellular osmotic pressure increases with the increase of $Na^+$ inside the cell. As a result, fluid enters the erythrocytes and becomes spherical. This mechanism is caused by the absence of spectrin from the proteins on the surface of erythrocytes and a decrease in the amount of lipids. The breakdown of erythrocytes is related to the specificity



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of blood circulation in the spleen. In the red pulp, part of the blood goes out of the sinuses, that is, into the intersinus space. Here, erythrocytes fall into an environment with low glucose and cholesterol levels. Such an environment is considered unfavorable for erythrocytes, causing erythrocytes to suffocate and lose their elasticity. Therefore, erythrocytes can pass through the narrow sinuses of the spleen due to their deformation properties. The erythrocyte membrane can lose a certain part of its surface during the passage through the narrow intersinusoidal slit. If hemolysis does not occur, after the membrane defect disappears, erythrocytes shrink and return to the bloodstream. This is how microcytosis develops. During retransmission from the splenic sinuses, microspherocytes are engulfed by the macrophage system or are degraded without their participation. That is why splenectomy helps in this disease.

Also to hereditary membranopathies:

- ✓ Hereditary elleptocytosis
- ✓ Hereditary poikilocytosis
- ✓ Hereditary stomatosis
- ✓ Hereditary acanthocytosis
- ✓ Hereditary echinocytosis

Fermentopathy is an example of anemia caused by deficiency of the enzyme glucose-6-phosphate dehydrogenase. The disease is inherited in a dominant way linked to the X chromosome. Dominant anemia is rarely observed. It is known that the disease is manifested by hemolytic crises after the use of some drugs: sulfonamides, anti-malarial and anti-tuberculosis preparations. The above-mentioned drugs oxidize hemoglobin and stop its respiratory function. This condition is not observed in healthy people because they have an antioxidant system. The main component of the antioxidant system is the regeneration of glutathione, and the amount of regenerated glutathione decreases in glucose-6-phosphate dehydrogenase deficiency. Therefore, in these patients, the therapeutic dose of medicinal preparations with oxidizing properties oxidizes and breaks down hemoglobin. In the hemoglobin molecule, the heme is cut off and the globin chain precipitates, which is called a Gains body. These inclusions are eliminated by the spleen. In some cases, hemolytic crises occur when the Konsky bob plant is used or when flower pollen is smelled. Its active factors (vicin, convicin) reduce the power of the antioxidant system by oxidizing regenerated glutathione.

The most common of the hemoglobinopathies is sickle cell anemia. In such patients, instead of Hb A, Hb C is synthesized. Hereditary defect occurs due to replacement of glutathione with valine at position 6 of beta chain in Hb S. As a result, in hypoxia, the solubility of hemoglobin is sharply reduced. Hb C, which is 100 times less soluble than oxidized Hb and 50 times less soluble than Hb A, precipitates in the form of crystals in an acidic environment, deforms erythrocytes and gives them a sickle shape. Such erythrocyte membrane loses its resistance and undergoes hemolysis inside the vessel.

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