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THE SYNDROME OF UNFORTUNATE CONSEQUENCES HELPPA

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Annotation. *HELLP syndrome includes hemolysis (H-hemoIisis), increased activity of liver enzymes (el - elevated liver enzymes) and a decrease in platelet count (Ip - low playlet count), usually manifests itself as a complication of preeclampsia, but can also develop independently. HELLP syndrome develops with a frequency of 1-6 cases per 1000 pregnancies and in 4-12% of patients with severe preeclampsia. Preeclampsia is characterized hypertension, proteinuria and edema in the second or third trimester of pregnancy and develops in 5-7% of pregnant women. In 70% of cases, HELLP syndrome develops before childbirth, in 30% - in the postpartum period.*

Keywords*: HELLP syndrome, eclampsia, antiphospholipid syndrome, hemolysis, thrombocytopenia.*

HELLP syndrome in pregnant women with gestosis is found in 0,5–1% of cases, characterized by high maternal and perinatal mortality. The pathophysiology of this syndrome has not been sufficiently studied. Endothelial dysfunction is considered to be a key link in the pathogenesis. Damage to the endothelium and inflammatory reaction leads to the development of coagulopathy, increased platelet consumption and the formation of platelet-fibrin microthrombus.

Every year, from 550 to 600 thousand women die from causes associated with pregnancy and childbirth.

One of these pathological conditions is the so-called HELLP syndrome, described by L. Weinstein in 1982 [1], which is considered as one of the forms of preeclampsia and eclampsia, accounting for 10-12% of maternal mortality [2]. The term is an abbreviation of English words that describe the main manifestations of this syndrome - Hemolysis, Elevated Liver enzymes, and Low Platelets.

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The syndrome can develop both during pregnancy (70% of cases) and after childbirth (30%) [3]. In typical cases, it occurs in multiparous women with a burdened obstetric history over the age of 25 years. Available unrecognized obstetric sepsis [4] and some other pathological conditions.

Characteristic early clinical manifestations of the syndrome are nausea and vomiting (86%), pain in the epigastric region (86%) and right hypochondrium, severe swelling (67%) [4]. Pain in the epigastric region can lead to erroneous diagnosis of acute surgical pathology.

The key criteria for diagnosing HELLP syndrome are the presence of hemolytic anemia, lactate dehydrogenase activity in the blood serum more than 600 U/L (as an indicator of hemolysis), AST >70 U/L and blood platelet content less than 100·109/L [5], although unexpectedly, clinical symptoms such as headache, visual disturbances, epigastric pain, nausea and vomiting, turn out to be more accurate criteria for the unfavorable outcome of the syndrome than laboratory parameters [6].

The pathogenesis of HELLP syndrome is not well understood. We can definitely say that there is a genetic predisposition to it, as evidenced by both its reoccurrence in the same woman during the next pregnancy and family history.

There are 178 genes that, according to the literature, are related to the development of preeclampsia and HELLP syndrome [7]. It is believed that the occurrence of the syndrome is based on an autoimmune reaction, as evidenced by the presence of antiplatelet, antiendothelial and other autoantibodies in the serum of such patients [4]. A number of authors, not without reason, draw a parallel between pregnancy and allotransplantation.

Since trophoblast invasion brings fetal tissue into contact with maternal immunocompetent cells, it is accompanied by an appropriate immune response to fetal soluble HLA antigen (sHLA-DR). Due to the fact that this antigen is capable of inducing apoptosis, even small concentrations of it can affect the immune system of the maternal body and the immune balance between it and the fetal body [8

These changes occur in the first trimester of pregnancy [9].

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With HELLP syndrome, high concentrations of the sHLA-DR antigen are always detected in the blood of pregnant women, and therefore its presence can be considered an acute reaction of transplant (fetal) rejection. Moreover, detection of this antigen in the blood of pregnant women with preeclampsia can be used to determine their risk of developing HELLP syndrome [8].

Differential diagnosis of HELLP syndrome is very difficult. Diseases with which it is necessary to differentiate HELLP syndrome include [11]:

1. Placental abruption.
2. Acute fatty hepatosis of pregnant women.
3. Anemia and thrombocytopenia.
4. Antiphospholipid syndrome in combination with pregnancy.
5. Eclampsia.
6. Hemolytic anemia.
7. Hemolytic Uremic syndrome.
8. Vomiting of pregnant women..
9. Hypertension and the occurrence of concomitant pregnancy.
10. Nephrolithiasis.
11. Peptic ulcer.
12. Preeclampsia.
13. Thrombocytopenia in pregnant women.
14. Thrombotic thrombocytopenic purpura.
15. Viral hepatitis.
16. Cytomegalovirus infection and infectious mononucleosis [1].

Cocaine addiction [1]. Immediate delivery is indicated for women with HELLP syndrome after 34 weeks' gestation. During labor and for 24 hours after delivery, patients should receive intravenous magnesium sulfate to prevent eclampsia, usually given as a loading dose of 4 grams followed by 2 g/hour. If the patient is already in labor, vaginal delivery is possible in the absence of signs of fetal distress or disseminated intravascular coagulation.

For any symptom of the development of multiple organ dysfunction, renal failure, or placental abruption, delivery should be performed immediately, usually by cesarean section. Induction of labor is not indicated for these patients, as this process can take from several hours to several days and pose a threat to the mother and fetus. Platelets are typically transfused if the platelet count is less than 20,000/mm3, or if a caesarean section is necessary if the platelet count is less than 50,000/mm3, or if there is any significant bleeding. Multiple platelet transfusions are usually not required in the absence of significant bleeding, as delivery will eventually resolve the thrombocytopenia

Forecast. HELLP syndrome is associated with an increased risk of maternal and fetal morbidity and mortality. The risk of maternal mortality is approximately 1%. HELLP syndrome is associated with multiple maternal complications, including pulmonary edema, acute renal failure, disseminated intravascular coagulation, placental abruption, liver hemorrhage or liver failure, and acute respiratory distress-syndrome and stroke. Patients with HELLP syndrome also have a higher risk of blood transfusions.

HELLP syndrome not only increases maternal morbidity and mortality, but also fetal morbidity and mortality. The incidence of perinatal mortality ranges from 7.4 to 20.4% and is largely dependent on gestational age and any additional complicating factors during pregnancy or childbirth.

The highest rates of fetal morbidity and mortality associated with early gestation (<28 weeks) are no higher than the rates of morbidity and mortality for fetuses of the same gestational age in women diagnosed with preeclampsia only.

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