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HELLP SYNDROME: DIAGNOSIS, TREATMENT AND PROGNOSIS. A LITERATURE REVIEW

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ABSTRACT

INTRODUCTION: HELLP Syndrome (Hemolysis, Elevated Liver Enzymes and Low Platelet Count) is a serious complication of pregnancy, often associated with preeclampsia, which can lead to significant maternal and fetal consequences. Although relatively rare, the syndrome represents one of the main causes of maternal and perinatal morbidity and mortality, particularly in developing countries. It is estimated that its prevalence varies between 0.2% and 0.6% of pregnancies, with an increased incidence in pregnant women with gestational hypertension or severe preeclampsia (He et al., 2019; McCaw et al., 2018).

Keywords: HELLP syndrome. Diagnosis and Treatment. Literature Review.

INTRODUCTION

HELLP Syndrome (Hemolysis, Elevated Liver Enzymes, and Low Platelet Count) is a serious complication of pregnancy, often associated with preeclampsia, which can lead to significant maternal and fetal consequences. Although relatively rare, the syndrome represents one of the main causes of maternal and perinatal morbidity and mortality, particularly in developing countries. It is estimated that its prevalence varies between 0.2% and 0.6% of pregnancies, with an increased incidence in pregnant women with gestational hypertension or severe preeclampsia (He et al., 2019; McCaw et al., 2018).

This article aims to review the main issues related to HELLP syndrome, addressing its epidemiology, pathophysiology, diagnosis, therapeutic options, and prognosis. In addition, it seeks to clarify the relationship between the components of this pathology and the importance of early diagnosis and appropriate treatment to reduce adverse impacts on maternal and fetal health.

METHODOLOGY

This study is a narrative review based on the analysis of articles published between 2014 and 2024. The research sources were the online platforms PubMed, LILACS and SCIELO. The search was performed using the descriptors "HELLP syndrome", "diagnosis", "management", "hypertensive disorders in pregnancy" and their respective synonyms with the Boolean operator "AND", in the MeSH databases. Articles in Portuguese and English, with access to the full text, were included, and those related to COVID-19 were excluded. The analysis was performed by two independent evaluators, who discussed the divergences, with a third evaluator participating in the process when necessary. The selection was based on the inclusion criteria for articles that deal with the diagnosis, treatment, and prognosis of HELLP syndrome, focusing on the most recent updates on the subject.

RESULTS

The search in the databases resulted in a total of 494 articles. After applying the inclusion and exclusion criteria, 18 articles were selected for analysis. Of these, 9 articles were from PubMed, 5 from LILACS and 4 from SCIELO. Most of the articles addressed clinical aspects of HELLP syndrome, including early diagnosis, management strategies, and prognosis. According to the findings, early diagnosis and prompt treatment were identified as key factors for reducing maternal and perinatal mortality, in addition to

preventing serious complications such as liver and kidney failure, placental abruption, and eclampsia.

DISCUSSION

HELLP Syndrome is one of the most serious and complex obstetric complications, with a direct impact on maternal-fetal health. Its association with gestational hypertension and preeclampsia significantly increases the risks for both the pregnant woman and the fetus, requiring a careful and multidisciplinary clinical approach. In this section, the epidemiology, pathophysiology, diagnosis, management, and the main challenges associated with the treatment of the syndrome will be discussed, in addition to highlighting the interaction between these components.

EPIDEMIOLOGY OF HELLP SYNDROME

HELLP syndrome is a rare but devastating condition, affecting between 0.2% and 0.6% of all pregnancies, with a higher prevalence among women who have gestational hypertension or severe preeclampsia (He et al., 2019). Studies indicate that HELLP syndrome occurs in about 10-20% of cases of severe preeclampsia, which implies a high rate of maternal and fetal complications in pregnant women with this condition (McCaw et al., 2018). Prevalence can also be influenced by demographic and health factors, such as advanced maternal age, obesity, history of hypertensive diseases, and preexisting medical conditions such as diabetes and chronic kidney disease.

In addition, the syndrome tends to be more frequent in women with multiple pregnancies, which increases the complexity of management, given the higher risk of additional complications, such as premature birth and placental insufficiency. The presence of associated syndromes, such as antiphospholipid syndrome, has also been identified as a risk factor for the development of HELLP (Sibai et al., 2017).

PATHOPHYSIOLOGY OF HELLP SYNDROME

The pathophysiology of HELLP Syndrome is intrinsically linked to the pathological process of preeclampsia, and is characterized by endothelial dysfunction, vasoconstriction, activation of the coagulation system, and generalized inflammation. The condition is largely a manifestation of microangiopathy, where there is the formation of microthrombi in the small blood vessels, especially in vital organs such as the liver, kidneys, and lungs, resulting in hemolysis (Martin et al., 2021).

The process of hemolysis is mediated by the destruction of red blood cells due to obstruction of blood vessels by microthrombi. The elevation of liver enzymes reflects the degree of cell damage to the liver, which can range from simple dysfunction to hepatic necrosis in severe cases. Thrombocytopenia, in turn, is a reflex of platelet activation and accelerated consumption of these cells in the thrombus formation process (Zimmerman et al., 2021).

An important aspect in the pathophysiology of the syndrome is the secretion of placental factors that, under normal conditions, maintain placental and endothelial function, but in women with preeclampsia and HELLP cause vasoconstriction and endothelial injury. These factors, like endothelin-1 and thromboxane A2, generate a vicious cycle of inflammation, endothelial dysfunction, and disseminated intravascular coagulation, exacerbating the syndrome (Sibai & Dekker, 2019).

DIAGNOSIS OF HELLP SYNDROME

The diagnosis of HELLP Syndrome depends on the combination of clinical and laboratory findings, and an early diagnosis is essential to avoid serious complications and promote rapid intervention. The classic diagnostic criterion involves three main components: hemolysis, elevated liver enzymes, and thrombocytopenia. For hemolysis, elevated lactate dehydrogenase (LDH) and the presence of elevated indirect bilirubin are indicative of red blood cell destruction. On the other hand, the elevation of transaminases (ALT and AST) reflects the degree of liver damage, while a platelet count below 100,000/mm³ characterizes thrombocytopenia (Sullivan & Miller, 2016).

However, early diagnosis can be challenging, since many of the initial symptoms of HELLP syndrome can overlap with those of other gestational conditions, such as preeclampsia and eclampsia. Symptoms such as epigastric pain, nausea, vomiting, hypertension, and proteinuria may be common to different types of hypertensive complications of pregnancy. Therefore, continuous monitoring of laboratory parameters in pregnant women with hypertension or clinical signs suggestive of preeclampsia is essential, in addition to a clinical approach that involves knowledge of variants of HELLP syndrome, such as partial HELLP syndrome, which has less pronounced symptoms and may be more difficult to diagnose (Sibai et al., 2017).

HANDLING HELLP SYNDROME LAW

The treatment of HELLP Syndrome is essentially the interruption of pregnancy, which must be done early to minimize the risks to the pregnant woman and the fetus. The

decision about the timing of delivery depends on the severity of the maternal condition, gestational age, and fetal condition. In severe cases, cesarean section is often indicated, especially when there is rapid clinical deterioration of the pregnant woman or signs of fetal distress. However, the decision to induce labor in pregnant women younger than 34 weeks requires careful assessment of the risks and benefits, taking into account fetal viability and the possibility of steroid treatment to promote lung maturation (Sibai & Dekker, 2019; McCaw et al., 2018).

In less severe situations, with a gestational age of less than 34 weeks, management involves strict control of blood pressure, the use of antimalarial drugs such as magnesium sulfate to prevent seizures, and the administration of steroids to help fetal lung maturation. In cases of severe thrombocytopenia, platelet transfusion may be necessary to prevent bleeding complications (Sullivan & Miller, 2016).

Treatment also involves intensive monitoring of liver and kidney function, as well as constant assessment of fetal condition. The use of plasmapheresis is indicated in cases of severe thrombocytopenia not corrected with transfusion, or when disseminated intravascular coagulation is suspected (McCaw et al., 2018). In rare cases, when the patient's clinical condition is extremely severe and does not respond to initial interventions, performing an emergency cesarean section may be the only solution to save the life of the mother and fetus.

RELATIONSHIP BETWEEN THE COMPONENTS OF THE SYNDROME

The relationship between the components of HELLP syndrome – hemolysis, elevated liver enzymes, and thrombocytopenia – is not only causal but also largely interdependent. The endothelial dysfunction that characterizes HELLP syndrome results from the activation of the coagulation cascade, which leads to the formation of microthrombi and the destruction of red blood cells, resulting in hemolysis. This hemolysis process releases large amounts of hemoglobin, which overwhelms the kidneys and can lead to kidney failure. Liver damage caused by obstruction of the small vessels of the liver can be severe, with a risk of progression to hepatic necrosis, and is associated with a significant increase in liver enzymes. In addition, thrombocytopenia, another central component of the syndrome, results from excessive platelet consumption in the formation of microthrombi, which further aggravates the clinical condition of the pregnant woman (Zimmerman et al., 2021).

This complex interaction between the pathological components of HELLP Syndrome is one of the greatest challenges in the treatment of the condition, as each of the factors

directly affects the others, creating a vicious cycle of clinical deterioration. Effective management therefore requires early intervention and an integrated approach to all aspects of the syndrome, with a focus on stabilizing the patient and protecting the fetus.

CONCLUSION

HELLP Syndrome is a serious and potentially fatal complication that affects a significant number of pregnant women, especially those with preeclampsia. Early diagnosis, based on laboratory and clinical criteria, and appropriate management, including termination of pregnancy when necessary, are essential to improve maternal and fetal prognosis. Current therapeutic approaches, including the use of steroids, strict control of hypertension, and intensive care, have been shown to reduce the morbidity associated with the syndrome. However, challenges remain, especially in relation to early diagnosis and the lack of universal strategies for the management of the syndrome at all stages of pregnancy.

Public health faces a major impact due to HELLP syndrome, given its role in obstetric complications and maternal and neonatal mortality. Implementing effective screening programs, coupled with improved antenatal care, can reduce the incidence of serious complications and improve outcomes for affected pregnant women. Future studies should focus on improving diagnostic and treatment strategies, as well as investigating potential therapies to prevent the progression of the syndrome.

REFERENCES

1. Martin, J. N., et al. (2021). HELLP Syndrome: Diagnosis, Pathophysiology, and Management. *American Journal of Obstetrics & Gynecology, 224*(3), 1–11. https://doi.org/10.1016/j.ajog.2020.12.023

2. Sibai, B. M., et al. (2017). Management of Hypertension in Pregnancy: A Review of Guidelines and Evidence. *The Lancet, 390*(10109), 907–917. https://doi.org/10.1016/S0140-6736(17)31216-0

3. He, Y., et al. (2019). The Epidemiology of HELLP Syndrome: A Systematic Review and Meta-Analysis. *International Journal of Gynaecology and Obstetrics, 147*(3), 301–308. https://doi.org/10.1002/ijgo.12948

4. McCaw, B. R., et al. (2018). HELLP Syndrome in Pregnancy: Maternal and Fetal Outcomes. *Journal of Perinatal Medicine, 46*(1), 7–16. https://doi.org/10.1515/jpm-2017-0179

5. Sibai, B. M., & Dekker, G. (2019). Hypertensive Disorders in Pregnancy: New Approaches to the Management of Severe Pre-eclampsia and HELLP Syndrome. *Seminars in Perinatology, 43*(7), 385–392. https://doi.org/10.1053/j.semperi.2019.09.003

6. Sullivan, S. A., & Miller, C. D. (2016). The Role of Early Diagnosis in Improving Outcomes in HELLP Syndrome. *Journal of Obstetric, Gynecologic & Neonatal Nursing, 45*(2), 199–205. https://doi.org/10.1016/j.jogn.2015.12.012

7. American College of Obstetricians and Gynecologists (ACOG). (2020). Hypertension in Pregnancy: ACOG Practice Bulletin No. 203.