



THROMBOCYTOPENIA AND HEMOGLOBINOPATHIES IN PREGNANCY

Thrombocytopenia and hemoglobinopathies are two hematological disorders that may be seen in pregnancy. These blood disorders are fairly uncommon, but can cause serious complications.

Q. WHAT IS THROMBOCYTOPENIA?

A. Thrombocytopenia is defined as a low platelet count. When the maternal platelet count falls below 150,000, thrombocytopenia is thought to be present. Several consequences can result from low platelet counts, specifically bleeding. It has been shown that difficult-to-control bleeding during surgery often results when platelet counts are 50,000 or less. When platelet counts are 20,000 or less, spontaneous bleeding has been reported.

Q. WHAT IS A HEMOGLOBINOPATHY?

A. Hemoglobinopathies are caused by abnormalities in the structural components of the hemoglobin chain. Because of the abnormal hemoglobin, anemia and inadequate transfer of oxygen ensues. The most common hemoglobinopathies are sickle cell disease, sickle cell trait, hemoglobin C disease, hemoglobin SC disease, and thalassemias.

Sickle cell disease is a chronic condition that results in the formation of abnormal red blood cells. It is the result of abnormalities in the chains that make up hemoglobin. People with this disease may have complications including increased risks of infection, strokes, bone damage, kidney damage, lung damage, spleen damage, eye damage, and stunted growth. These patients have a shorter life expectancy as well.

Sickle cell trait is a benign disorder. Few complications exist with sickle cell trait. Of importance is the sickle cell status of the father of the baby. If the father is sickle cell trait positive, the infant has a 25% risk of having sickle cell disease.

Hemoglobin C and SC diseases are similar to sickle cell disease, but are a lesser form. These patients, however, are followed closely in pregnancy, as are the sickle cell patients.

Thalassemias can cause significant hematologic problems in pregnancy. These disorders result from abnormal rates of formation of the chains that make up hemoglobin. There are several different types of thalassemia, and some are severe and life-threatening, while other forms are mild in nature. These disorders are

genetic, and the incidence has been reported at about 1 in 500. Ethnic groups thought to be at risk of thalassemias include northern Europeans, Japanese, Eskimo, and Native American backgrounds. As with sickle cell disease, patients with thalassemias are followed closely during pregnancy. Additionally, the prenatal diagnosis of thalassemia is often attempted in an effort to give parents information of the possibility of this disease in their child.

Q. HOW DOES THROMBOCYTOPENIA AFFECT PREGNANCY?

A. Four percent of pregnant patients have what is known as gestational thrombocytopenia. This is a benign condition seen only in pregnancy that has no effect on the fetus and usually resolves with delivery. With gestational thrombocytopenia, platelet count rarely falls below 100,000. If the platelet count falls below 75,000, the physician must rule out other potential causes of the thrombocytopenia.

The most common causes of thrombocytopenia in pregnancy are ITP (idiopathic thrombocytopenic purpura), thrombocytopenia with preeclampsia (a hypertensive disorder of pregnancy), and gestational thrombocytopenia.

Idiopathic thrombocytopenic purpura (ITP) is an autoimmune disorder. It affects approximately 1 to 3 per 1000 pregnancies. In this condition, the mother produces antibodies to her own platelets. This condition has not been shown to be caused by pregnancy or to change in severity with pregnancy. The most important aspect of ITP in pregnancy is the potential passage of maternal antibodies across the placenta to the baby. Because of this, the baby may have profound thrombocytopenia at birth. This thrombocytopenia in the baby may lead to significant bleeding with the largest concern being for bleeding in the head.

Thrombocytopenia associated with preeclampsia can be a serious condition as well. Preeclampsia is a disease seen only in pregnancy. It was known as toxemia in the past and is characterized by elevated blood pressures, protein in the urine, and nondependent edema (swelling). A severe form of preeclampsia can occur known as HELLP syndrome (an acronym standing for hemolysis, elevated liver function tests, and low platelets). With HELLP syndrome, a woman can become severely ill. The treatment of HELLP syndrome is delivery of the infant.

Finally, gestational thrombocytopenia is as mentioned above.

Q. HOW DOES A HEMOGLOBINOPATHY AFFECT PREGNANCY?

A. As mentioned above, there are several different types of hemoglobinopathies, and each of these can have an effect on pregnancy. In the mother, the most profound disorders result in anemia. For sickle cell anemia, the mother must be followed closely in pregnancy because of the increased risk of infections and sickle cell crisis. For the thalassemias, a similar clinical course in pregnancy is often seen. If a woman has a severe form of thalassemia, however, she may not live to childbearing age.

Of greatest importance in the hemoglobinopathies and pregnancy is the need to offer couples with the genetic disorders or who are carriers of the genetic disorders

prenatal diagnostic testing for their infant. Often a sample of their infant's blood can be obtained by a specialist in high-risk pregnancies, allowing them to diagnose a hemoglobinopathy in their child prior to birth.

Q. HOW IS THROMBOCYTOPENIA TREATED?

A. No treatment is necessary for gestational thrombocytopenia. For ITP, treatments of maternal thrombocytopenia include glucocorticoid therapy, platelet transfusions as indicated, gamma globulin therapy, or splenectomy. If the fetus is found to have thrombocytopenia with platelet counts less than 50,000, the risks and benefits of vaginal delivery and cesarean section will likely be discussed with your physician.

Q. HOW IS A HEMOGLOBINOPATHY TREATED?

A. Treatment of hemoglobinopathies in pregnancy varies depending on the symptoms. Because of baseline low hemoglobin levels, women are often supplemented with folic acid during pregnancy. They are followed closely for signs of infection, specifically urinary tract infections. If you have a hemoglobinopathy, your fetus will be monitored closely during the latter part of pregnancy with serial ultrasound studies and monitoring to assure the well being of your baby. Finally, if your hemoglobin level drops very low in labor and delivery, your physician may recommend a blood transfusion.

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