



SICKLE CELL DISEASE IN PREGNANCY

Sickle cell disease is a disorder seen primarily in African Americans. Due to a genetic substitution of DNA from the hemoglobin chain (the chain that makes up red blood cells), abnormal red blood cells are produced that change shape and "sickle." Approximately 1 in 12 African-American people has one gene for sickle cell disease and therefore has what is called sickle cell trait. Approximately 1 in 576 African-American people have sickle cell disease. It is estimated that over 2.5 million Americans have sickle cell trait, with over 50,000 with sickle cell disease. Over 1000 infants are born with sickle cell disease each year. The mean age of death for a woman with sickle cell disease is 48 years.

Q. WHAT IS SICKLE CELL DISEASE?

A. Sickle cell is a disease of red blood cells. It is an inherited disorder that involves abnormal hemoglobin (the main substance in a red blood cell that is responsible for the transfer of oxygen). Normal red blood cells contain hemoglobin A. In sickle cell disease the red blood cells contain mostly hemoglobin S, an abnormal form. Cells with this abnormal form of hemoglobin often change shape from a normal round shape, to a crescent shape (sickled shape), hence the name sickle cell anemia. The sickle shaped cells often get caught in small blood vessels, decreasing the blood flow and oxygenation of certain organs, as well as causing pain. Additionally, they often break apart as they go through these small vessels, decreasing the total amount of red blood cells in the body with resultant anemia (low red blood cell count). Of additional importance is the fact that the normal life span of a red blood cell is approximately 120 days, whereas that of a sickled cell is only 5 to 10 days.

Several complications can occur in a person who has sickle cell disease. These complications are a direct result of blockage of blood flow and breaking up of sickled red blood cells. They include episodes of pain crisis which can last from a few days to several weeks, increased risk of infections, increased risk of strokes, bone damage, kidney damage, lung damage, spleen damage, liver damage, eye damage, anemia (low red blood cell counts), and stunted growth.

Q. HOW IS SICKLE CELL DISEASE DIAGNOSED?

A. Sickle cell disease is an inherited disorder. In the United States it is primarily seen in people of African American descent, but it is also seen in Middle Eastern, Indian, and Mediterranean individuals. Diagnosis is made by a blood test known as a hemoglobin electrophoresis. This test isolates the different hemoglobin chains, and can determine if you have the normal hemoglobin A, abnormal hemoglobin S, or a combination of both which results in sickle cell trait. It has now become fairly standard to test newborn infants for sickle cell disease.

Q. HOW IS SICKLE CELL DISEASE TREATED?

A. There is no true treatment for sickle cell disease. All that can be done is to attempt to decrease the risk of any of the serious complications of sickle cell disease, and improve quality of life. Of most importance in this respect, is attempting to prevent serious infection. It is imperative that all sickle cell patients receive the recommended childhood immunizations in a timely fashion. Additionally, patients with sickle cell disease should receive a pneumococcal vaccine, since they are at increased risk of pneumonia, especially if they have spleen damage. These patients should take daily vitamins with folate to help with red blood cell production. They should get plenty of rest, and avoid extremely stressful situations or situations requiring extreme exertion since both of these can put increased demand on their bodies for oxygen. Sickle cell patients should drink at least 8 8-ounce glasses of water a day in an attempt to keep well hydrated, and hopefully decrease the blockage of the small blood vessels by the sickled cells. If any unusual signs or symptoms appear, or if any signs of infection appear (such as fever, pain, etc.) patients with sickle cell disease should see their doctor immediately.

Q. HOW DOES SICKLE CELL DISEASE AFFECT PREGNANCY?

A. Pregnancy is a significant stress for a woman with sickle cell disease. Because of this we see more pain crisis episodes, increased risk of infection, and worsening anemia. We also see increased risk of miscarriage, stillbirth, or neonatal death (death around the time of delivery) in patients with sickle cell disease. The incidence of this approaches 30%. Studies have shown an increased risk of maternal death associated with sickle cell disease.

Q. HOW IS SICKLE CELL DISEASE TREATED IN PREGNANCY?

A. All people who fit the demographic description of being at high risk for sickle cell disease should be screened for the disease during pregnancy. Patients with positive screens should undergo hemoglobin electrophoresis to determine if they have sickle cell disease. If a woman has sickle cell trait, she is not at risk of having adverse outcomes during pregnancy. Her partner, however, should be tested as well. If he has either sickle cell disease or sickle cell trait, the couple will be at risk of having an infant with sickle cell disease and should have prenatal counseling/diagnosis regarding this.

Because of the potential serious complications seen with sickle cell disease and pregnancy, your physician will follow you very closely during your pregnancy and may even refer you to a high risk specialist. Because of baseline low hemoglobin levels and increased need for red blood cell function in pregnancy, pregnant women require increased folic acid supplementation. Your physician may recommend approximately 1 milligram of folic acid a day.

Pregnant women are extremely susceptible to infection, with serious resultant complications. Asymptomatic bacteriuria (bacteria in the urine that causes no symptoms of a urinary tract infection) can be extremely dangerous for the pregnant patient with sickle cell disease. If left untreated this can quickly progress to a urinary tract infection and/or a kidney infection (pyelonephritis) with severe

consequences. Your physician will regularly screen your urine for asymptomatic bacteriuria.

Increased pain episodes are common in pregnancy, and are often precipitated by dehydration or infection. If you have a significant pain crisis, your physician will most likely admit you to the hospital. He or she will give you oxygen, intravenous fluids, and intravenous pain medications. The physician will then look for any sources of infection that may be attributable to the pain crisis.

Your physician will monitor fetal health and development very closely if you have sickle cell disease. Due to a high incidence of growth restriction of fetuses and a high risk of perinatal death (fetal death before or around the time of delivery), your physician may recommend ultrasounds every 3 to 4 weeks late in your pregnancy to assess the growth of your fetus. He or she may suggest routinely monitoring your baby either with fetal heart rate monitoring (known as a nonstress test or NST) or ultrasound monitoring (known as a biophysical profile or BPP).

Finally, during labor and delivery you will be monitored very closely. Epidural analgesia has been shown to be safe during pregnancy. If you require a cesarean delivery and your hemoglobin value is less than 7 gm/dl your physician may request that you have a blood transfusion prior to your surgery.

There is currently a debate over the benefit of prophylactic blood transfusions in pregnancy (giving a blood transfusion before any complication, to increase the blood count and attempt to prevent a complication). Your physician may discuss this as an option.

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