Sickle Cell Anaemia in Pregnancy: A Review

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ABSTRACT

Sickle cell anaemia is one of the groups of inherited disorders known as sickle cell disease and it affects the shape of red blood cells, which carry oxygen to all parts of the body. Sickle cell anaemia (SCA) is caused by a homozygous mutation (haemoglobin S) and presents as chronic anaemia accompanied by painful episodes. When someone carries the sickle cell trait, seeing a genetic counselor before trying to conceive can help her understand the risk of having a child with sickle cell anaemia. Women suffering from sickle cell anaemia are able to get pregnant normally. However, it is important to consult with the doctor enough time beforehand in their pregnancy plans so as to reduce the possibility of complications.

Keywords: Anaemia, Haemoglobin S, Pregnancy, Sickle Cell Anemia, Sickle Cell Trait.

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Introduction

Sickle cell anemia is one of the groups of inherited disorders known as sickle cell disease and it affects the shape of red blood cells, which carry oxygen to all parts of the body.[1-5] In sickle cell anaemia, some red blood cells are shaped like sickles or crescent moons. These sickle cells also become rigid and sticky, which can slow or block blood flow.[6]

Strictly, sickle cell anaemia (SCA) is caused by a homozygous mutation (haemoglobin S) and presents as chronic anaemia accompanied by painful episodes. The main defect triggering these events is impaired microcirculation due to sickling of erythrocytes.[7-12]

According to Koshy[13], Sickle cell disease refers to a group of genetic disorders characterized by the predominance of hemoglobin S. This includes sickle cell anemia (SS) sickle hemoglobin C disease (SC), sickle beta thalassemia plus (Sβ+ Thal), sickle beta thalassemia zero (β0Thal), sickle with alpha thalassemia (SSαThal) and rare combinations of sickle hemoglobin with Hb D, Hb O, etc.

According to Weatherall et al.[14], Sickle Cell Disease has its origins in sub-Saharan Africa and the Middle East, hence it is most prevalent in individuals of African descent as well as in the Caribbean, Middle East, parts of India and the Mediterranean, and South and Central America. Owing to population migration, SCD is now of increasing importance worldwide and there are increasing numbers of affected individuals in Europe and the United States of America (USA).

According to NHS Foundation Trust[15], the term Sickle Cell Disease (SCD) includes sickle cell anaemia (HbSS) and the heterozygous conditions of haemoglobin S and other clinically abnormal haemoglobins. These include a combination with haemoglobin C (giving HbSC), a combination with beta thalassaemia (giving HbSB thalassaemia), and a combination with haemoglobin D, E or O-Arab.

Sickle Cell Anemia in pregnancy

According to Muganyizi and Kidanto[16], Sickle Cell Anemia or Sickle Cell Disease in pregnancy is associated with increased adverse fetal and

maternal outcomes. In Tanzania where the frequency of sickle cell trait is 13% there has been scanty data on Sickle Cell Disease in pregnancy. In Tanzania for example, it is estimated that more than 11,000 babies are born with Sickle Cell Disease each year and without proper intervention, these children will die before reaching adulthood.[17] According to Muganyizi and Kidanto[16], the incidence of Sickle Cell Disease delivery at Muhimbili National Hospital (MNH) has been on the rise in the past 13 years. The disease is associated with excessive maternal deaths that are by far attributable to infections. Standard guidelines to improve the care of SCD in pregnancy are urgently needed in Tanzania.

Effect of sickle cell anaemia on maternal and fetal outcome

According to Dong et al.[18], Sickle cell anaemia (SCA) is a life-threatening genetic condition contributing to high-risk pregnancies affecting both the mother and fetus. With improved management of children with SCA, this life-threatening hematological disorder has evolved into a chronic disease of adults, and consequently, parenthood has now become a possible and important life goal for many patients.

Globally, around three-quarters of all those affected with sickle cell disease live in Sub-Saharan Africa, where an estimated 240 000 babies with HbSS are born every year. Nevertheless, in contrast to high-income regions, few studies have documented the natural history of sickle cell disease in Africa.[19]

Pregnancy complications. Sickle cell anemia can increase the risk of high blood pressure and blood clots during pregnancy. It can also increase the risk of miscarriage, premature birth, and having low birth weight babies (Mayo Foundation for Medical Education and Research, 2023).

According to Ware, de Montalembert et al.[20], repeated sickling and ongoing haemolytic anaemia, even when subclinical, lead to parenchymal injury and chronic organ damage, causing substantial morbidity and early mortality.

Although Sickle Cell Disease adversely affects pregnancy, leading to increased incidence of maternal and perinatal complications like pre-eclampsia, preterm labor, abortions, etc., adequate care throughout pregnancy ensures a better outcome.[7]

Management of sickle cell anemia in pregnancy

According to Mayo Foundation for Medical Education and Research[6], when one carries the sickle cell trait, seeing a genetic counselor before trying to conceive can help her understand the risk of having a child with sickle cell anemia. A genetic counselor can then explain possible treatments, preventive measures, and reproductive options.

With improved healthcare facilities, antibiotic prophylaxis, vaccination, and availability of drugs like hydroxyurea, Jain et al.[7] observed that the life expectancy of Sickle Cell Disease patients has improved and that more women are reaching the reproductive age group and are expressing their desire to reproduce.

According to Dong et al.[18], providing continuous management with healthy red blood cell function and avoiding Sickle cell anemia (SCA)-associated complications, such as pain crises, acute chest syndrome, and stroke, are crucial for a healthy pregnancy.

According to Ndeezi et al.[21], Uganda ought to include targeted neonatal screening in the districts with the highest-burden; training of healthcare providers, and establishment of regional and district sickle cell clinics to provide penicillin and antimalarial prophylaxis, family education, and immunisations; research of the safety, dosing, and benefits of hydroxyurea; and creation of a national sickle cell awareness strategy that will include premarital testing and counselling aimed at lowering the burden of sickle cell disease.

Conclusion

In conclusion, therefore, women suffering from sickle cell anemia are able to get pregnant normally. However, it is important to consult with

the doctor enough time beforehand in their pregnancy plans so as to reduce the possibility of complications. Furthermore, pregnant women suffering from sickle cell anemia are at a higher risk of giving birth to preterm babies or babies with low birth weight. However, most women are able to have healthy children if they are properly monitored and guided before, during, and after their pregnancies.

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