

Letter to the editor

Sickle Cell Disease in Pregnancy: Obstetrician's Nightmare in Resource-Poor Countries!

Boniface Uwaezuoke Odugu¹, Ifeanyi Johnson Onyekpa¹**How to cite:**

Odugu et al. 2023. "Sickle Cell Disease in Pregnancy: Obstetrician's Nightmare in Resource-Poor Countries!". *Journal of Applied Health Sciences and Medicine* 3(5): 32 – 35.

<https://doi.org/10.58614/jahsm358>

Received:

May 10, 2023

Accepted:

May 17, 2023

Published:

May 20, 2023

Corresponding author:

Odugu BU

boniface.odugu@esut.edu.ng

¹Department obstetrics and Gynaecology, Enugu State University of Science & Technology College of Medicine/Teaching Hospital, Enugu.

Sickle Cell Disease is a clinical condition in which the sickling gene occurs along with another abnormal gene affecting the quantity and quality of the haemoglobin [1]. It is a monogenic condition brought on by an A to T point mutation in the gene for β -globin, which results in aberrant Hb-S (Hb-S), which polymerizes in the deoxygenated state and causes physical distortion or sickling [2]. About 300,000 neonates worldwide are born each year with sickle cell disease, the most common hereditary hemoglobinopathy, with the majority being born in Nigeria, India, and the Democratic Republic of the Congo [3]. The term sickle cell disease includes different genotypes of homozygous HbS sickle cell anaemia (SS) and the double heterozygote states of sickle haemoglobin C disease (SC) sickle beta plus thalassaemia($S\beta$ +Thal), sickle beta zero thalassaemia ($S\beta$ 0thal), sickle cell anaemia with alpha thalassaemia ($SS\alpha$ thal) and sickle cell anaemia with high foetal haemoglobin (SS +F) [4]. The life expectancy of SCD patients has increased recently due to better medical facilities, antibiotic prophylaxis, vaccination against pneumococcal infections, accessibility to medications like hydroxyurea, and blood transfusion services. Female SCD patients in the reproductive age range are now expressing the desire to have children.

Recent advances in prenatal diagnosis and pre-implantation genetic diagnosis in Assisted Reproductive Technology (ART) help couples suffering from SCD to have a healthy baby but not without a myriad of challenges for the obstetrician; and this is the focus of this paper. SCD adversely affects pregnancy leading to increased incidence of maternal and perinatal complications like pre-eclampsia, preterm labour, IUGR abortions etc.

Preeclampsia, eclampsia, worsening vaso-occlusive crises, and acute chest syndromes are all rates of obstetric complications brought on by physiological changes in the circulatory, hematologic, renal, and pulmonary systems that occur during pregnancy. These changes can overburden organs that already have chronic injuries secondary to SCD. Additionally, placental vaso-occlusion causes villous fibrosis, necrosis, and infarction, which impairs uteroplacental circulation and results in prolonged foetal hypoxia and unfavourable foetal outcomes [5, 6].

In the beginning, both the mother's and the baby's pregnancy outcomes in SCD were dismal. However, due to better preconception, prenatal, delivery, and puerperal care in more developed climates, there has been a recent improvement. A maternal mortality rate of 0.38 - 1.29/100,000 births and a perinatal mortality rate of 1.21 - 2.50/100,000 births are still reported in sub-Saharan Africa, where the prevalence and complications



© 2023

by the authors. The terms and conditions of the Creative Commons Attribution (CC BY) licence apply to this open access article

of sickle cell disease are highest in the world. Unfortunately, fetomaternal outcomes have not improved significantly in this region [7].

Pre-eclampsia and eclampsia are obstetric problems that are more common in SCD patients than in the general population because pregnancy in the background of SCD is a high risk one [8].

The risk of gestational diabetes is also high [9], Microvascular damage and decreased utero-placental circulation in these mothers may lead to an exaggerated risk of spontaneous abortions and stillbirths. Other factors contributing to adverse foetal outcomes include poor general health of the mother and drug abuse like tobacco, alcohol, and narcotics [10]. Pregnancy exacerbates the pre-existing anaemia in SCD women, leading increases the level of lead in the blood during pregnancy. There is a higher rate of caesarean deliveries in SCD patients even though it is not an indication in itself [6] and does not come without its peculiar challenges and complications.

Infections like malaria, pneumonia, pyelonephritis, UTIs, and postpartum infections, among others, are more likely to occur when the immune system is already impaired during pregnancy and the defective splenic functions in SCD caused by auto-splenectomy are added to that condition. Predisposing SCD women to thromboembolic problems including deep vein and cerebral venous thromboses is pregnancy's hypercoagulable state. Maternal mortality is noticeably greater in SCD women compared to the general population as a result of a number of obstetric and non-obstetric problems. Maternal mortality has decreased in industrialised nations due to better healthcare infrastructure and increased awareness, but in underdeveloped nations like ours, the situation is still the same.

Pregnancy with sickle cell illness carries a substantial risk. This is crucial in West Africa, which is home to endemic malaria and is located in the sickle cell belt of the globe. Therefore, in such pregnancies, specialised prenatal, intrapartum, and post-natal care is indicated.

Painful crises, acute chest syndrome, pulmonary embolism, anaemia, and an increase in infections and infestations are among the majority of these difficulties. The common pain reliever acetaminophen is not always effective, and NSAIDs are not recommended between the 24th and 32nd week of pregnancy because they can cause premature closure of the ductus arteriosus. However, pain crises can happen at any stage of pregnancy. As analgesia is enhanced, the use of drugs can cause tolerance, dependence, addiction, and expense increases. The obstetrician faces these problems.

A clear diagnosis of one requires particular features on a chest x-ray (CXR), white cell counts, and D-dimer. Acute chest syndrome and pulmonary embolism may co-exist, making a diagnosis of one difficult. Leucocytosis is a common physiological sign, although CXR is often contraindicated during pregnancy. Therefore, patients who present with a cough, respiratory distress, or chest pain are typically treated based on their symptoms, which raises the cost of therapy. Obstetricians are faced with a problem when low molecular weight heparin is used to treat thrombo-embolism because it increases the risk of placental abruption.

Patients with SCD who are pregnant are more likely to contract infections such urinary tract infections (UTIs), respiratory tract infections (RTIs), hepatitis, transient bacteremia, osteomyelitis, and HIV—the latter of which is brought on by frequent

blood transfusions. SCD patients frequently experience acute cholecystitis, which can resemble other problems such as hepatopathy, hepatitis, and hepatic sequestration. Liver function tests that are not very specific during pregnancy may be needed for this diagnosis.

The problem of anaemia, which is made worse by physiological blood volume dilution during pregnancy, is of the utmost concern. Treatment for this illness is difficult since volume overload and an increased risk of blood-related infections, some of which could pose a public health danger like HIV, could result.

The first four days after delivery and the final four weeks of pregnancy are when these disorders are most common, hence foeto-maternal surveillance should be prioritised during this time. Additionally, the management of these patients during labour necessitates that the second stage be shortened, preferably using forceps, a skill that is currently on the decline among obstetricians due to a fear of legal action; additionally, caesarian delivery also has a high rate of anaesthetic complications among this subset of patients. The availability and affordability of epidural analgesia, which offers a great deal of hope during both vaginal and caesarian deliveries, are issues, particularly in the world's resource-poor nations.

Finally, managing sickle cell illness during pregnancy is extremely difficult for obstetricians. Understanding its aetiology, complications, and management restrictions may help us create a better care plan for our SCD pregnant women to improve the success of their pregnancies.

References

- [1] Kwawukume E.Y, Oppong S.A. (2015), Sickle cell disease in pregnancy in *Comprehensive Obstetrics in the tropics*; 2nd edition; pp 311-319.
- [2] Betty SP, Solomon F, Ofori-Acquah, Kenneth RP. Sickle cell disease: genetics, cellular and molecular mechanisms and therapies. *Anaemia*. 2012. Article ID 143594, 2 Pages, 2012. doi: 10.1016/0268-https://doi.org/10.1155/2012/143594
- [3] Piel FB, Hay SI, Gupta S, Weatherall DJ, Williams TN. Global burden of sickle cell anaemia in children under five, 2010-2050 modelling based on demographics, excess mortality, and interventions. *PloS Med*. 2013; 10(7):e1001484 doi: 10.1371/journal.pmed.1001484.
- [4] Koshy M. Sickle cell disease and pregnancy. *Blood Rev*. 1995;9(3):157-64. doi: 10.1016/0268-960X(95)90021-7.
- [5] Villers MS, Jamison MG, De Castro LM, James AH. Morbidity associated with sickle cell disease in pregnancy. *American Journal of Obstetrics and Gynecology*. 2008 Aug 1;199(2):125–e1. doi: 10.1016/j.ajog.2008.04.016.
- [6] Hassell K. Pregnancy and sickle cell disease. *Hematology/Oncology Clinics*. 2005 Oct 1;19(5):903–16. doi: 10.1016/j.hoc.2005.07.003.
- [7] Rahimy MC, Gangbo A, Adjou R, Dequenon C, Goussanou S, et al. Effect of active prenatal management on pregnancy outcome in sickle cell disease in an African setting. *Blood*. 2000;96:1685–1689.
- [8] Boafor TK, Olayemi E, Galadanci n, Hayfron-Benjamin C, Dei-Adomakoh Y, Segbefia C, Kassim AA, Aliyu MH, Galadanci H, Tuuli MG, Rodeghier M, Debaun MR. Pregnancy outcomes in women with sickle-cell disease in low and high income countries: a systematic review and meta-analysis. *BJOG*. 2016 Apr;123(5):691–8. doi: 10.1111/1471-0528.13786.
- [9] Barfield WD, Barradas DT, Manning SE, Kotelchuck M, Shapiro-Mendoza CK. Sickle cell disease and pregnancy outcomes: women of African descent. *Am J Prev Med*. 2010;38(4):S542–9. doi: 10.1016/j.amepre.2009.12.020.

- [10] Powars DR, Sanhu M, Niland-Weiss J, Johnson C, Bruce S, Maming PR. Pregnancy in sickle cell disease. *Obstet Gynecol.* 1986;67:217–228. doi: 10.1097/00006250-198602000-00012.