CASE REPORT

# Pregnancy in a Sickle Cell Disease Patient: A Nightmare!

Bhavya Doshi[1], Madhuri A Mehendale[2], Arun H Nayak[3], Archana A Bhosale[4], Snehal Mulik[5]

## Abstract

**Aim:** To discuss the effect of pregnancy in sickle cell disease (SCD) patients and its associated complications.

**Background:** Sickle cell disease is the most common inherited disorder worldwide and in certain regions of India with varying clinical severity and potentially serious complications. Sickle cell disease can magnify complications during pregnancy and in turn negatively influence the pregnancy outcomes. The physiological adaptations during pregnancy that occur in the circulatory, hematologic, renal, and pulmonary systems can overburden organs that already have chronic injuries secondary to SCD, thus increasing the rate of obstetric complications like miscarriage, anemia, preeclampsia, worsening of vaso-occlusive crisis, and acute chest syndromes.

**Case description:** A 23-year-old Indian primigravida patient, known case of SCD with anemia and splenic infarct with h/o multiple blood transfusions. The patient presented at 12 weeks with intrauterine fetal demise and was medically aborted. The post-abortion patient was posted for splenectomy as she had episodes of hemolytic jaundice. Post-splenectomy patient further developed bowel obstruction and thrombus formation in the infrarenal part of inferior vena cava (IVC). She was again operated and for obstruction and the band was removed. For thrombi, patient was given low molecular weight heparin (LMWH). The patient was finally discharged on tb. hydroxyurea and other antibiotics.

**Conclusion:** The higher rate of complications occurs in women with sickle cell crisis exaggerated by underlying factors such as long-term anemia and pregnancy increases the risk further. Thus, a multidisciplinary approach with regular follow-up of SCD patients since the time of preconceptional time is important to avoid pregnancy-related complications and also for a better pregnancy outcome.

**Clinical significance:** The physiological changes of pregnancy like increased blood volume, increased metabolic demand, increased blood viscosity, and hypercoagulability get aggravated in SCD patients leading to increased incidence of complications. Prepregnancy anemia and other complications of a mother can further affect the outcome, thus preconceptional counseling is a crucial part of management.

**Keywords:** Multidisciplinary approach, Pregnancy, Sickle cell disease.

*Journal of South Asian Federation of Obstetrics and Gynaecology* (2020): 10.5005/jp-journals-10006-1821

## Background

Sickle cell disease (SCD) is the most common hereditary disease worldwide and in certain regions of India with varying clinical severity and potentially serious complications.[1] Globally, India accounts for 14.5% of the total newborns with SCD.[1] Sickle cell disease can magnify complications during pregnancy and thus negatively influence pregnancy outcomes. The term SCD includes different genotypes of homozygous HbS sickle cell (SS) anemia and the double heterozygote states of sickle hemoglobin C disease (SC), sickle beta plus thalassemia (Sβ+Thal), sickle beta zero thalassemia (Sβ0thal), sickle cell anemia with alpha thalassemia (SS αthal), and sickle cell anemia with high fetal hemoglobin (SS+F).[2] Sickle cell anemia (SCA) is caused by a homozygous mutation (hemoglobin S) and presents as chronic anemia accompanied by painful episodes. The main defect triggering these events is impaired microcirculation due to sickling of erythrocytes.[2]

Previously, the management of patients with SCA was poor, and pregnancy was associated with high maternal and fetal mortality. Nowadays with newborn screening techniques, maternal screening, and preventive measures like vaccination and antibiotic prophylaxis since birth, overall disease outcomes and patient survival have improved and there is a significant reduction in maternal and neonatal mortality rates as well.[3] However, despite all advances, pregnancy in SCD is still associated with higher clinical and obstetric complications compared with the general population.

The physiological adaptations that occur in the circulatory, hematologic, renal, and pulmonary systems during pregnancy can overburden organs that already have chronic injuries secondary to SCD, increasing the rate of obstetric complications like anemia and preeclampsia, worsening of vaso-occlusive crisis, and acute chest syndromes. Microvascular damage and decreased uteroplacental circulation in these mothers leads to an exaggerated risk of spontaneous abortions and stillbirths.[4] Though pregnancy in SCD carries a greater of fetal and maternal complications, it can be managed by ensuring adequate preconceptional, antenatal, and perinatal care.

[1–5]Department of Obstetrics and Gynecology, Lokmanya Tilak Municipal General Hospital and Medical College, Mumbai, Maharashtra, India

**Corresponding Author:** Madhuri A Mehendale, Department of Obstetrics and Gynecology, Lokmanya Tilak Municipal General Hospital and Medical College, Mumbai, Maharashtra, India, Phone: +91 9769363190, e-mail: drmadhuri1982@yahoo.co.in

**How to cite this article:** Doshi B, Mehendale MA, Nayak AH, *et al.* Pregnancy in a Sickle Cell Disease Patient: A Nightmare!. J South Asian Feder Obst Gynae 2020;12(5):323–325.

**Source of support:** Nil

**Conflict of interest:** None

## Case Description

A 23-year-old Indian primigravida patient presented at 9 weeks of gestation with a complaint of severe pain in the abdomen, body ache for 1 day, and fever for 2 days. On examination, she had grade III pallor, icterus, splenomegaly, and tenderness on abdominal palpation. Her investigations read as hemoglobin (Hb)—6.7 g%,

white blood cells (WBCs)—3,800, platelet—88,000, T. bilirubin—9.3, contrast enhanced computed tomography (CECT) (abdomen + pelvis) s/o splenic infarct with other investigations being normal. The patient was transfused with two pints of a packed red cell after which her Hb was 8 g%. Her COVID swab was negative.

The patient was a diagnosed case SCD since 2016 with Hb electrophoresis s/o SS type SCD. She has had a history of intermittent blood transfusions since then. The last blood transfusion was 6 months ago. She had an episode of fever and breathlessness for which she was admitted to the medicine ward 4 months ago. She is on tb. hydroxyurea, tb. folic acid, tb. calcium, syrup sucralfate, and tb. cefixime since then. The patient was instructed to follow-up for splenectomy after few weeks.

Later the patient presented to us at 12 weeks of gestation with ultrasonography (USG) obstetric which was s/o intrauterine fetal demise of 12 weeks of gestation. Her prothrombin time (PT)/ International Normalized Ratio (INR) were 19.6/1.56, respectively. Her Hb was low and the SGOT value was deranged. She has transfused with 2-pint packed red blood cells (PRC) following which the patient went into the process of spontaneous abortion. She was aided in the process medically and her immediate postabortal period was uneventful. Post-aborting on day 1 patient c/o pain in abdomen, tenderness, and guarding. Her CT abdomen + pelvis was s/o early changes of bowel wall ischemia, moderate splenomegaly, multiple splenic infarcts. Her Hb was 5.9 g%, WBC—2,400, platelet—46,000. She was transfused with 3-pint PRC and 3-pint FDP. Total bilirubin—11.8 which was grossly elevated, serum glutamic-oxaloacetic transaminase (SGOT)/ serum glutamic pyruvic transaminase (SGPT)—528/82 which were deranged, blood urea nitrogen (BUN)/creatinine—24/0.7 which were normal, and PT/INR—22/1.95 were also deranged. The patient became drowsy on day 3 with pallor and icterus as positive clinical findings. She was shifted to Medical ICU for critical care management. The patient developed hematemesis, fever. On examination, bilateral crepitations were appreciated. She was kept on $O_2$ by mask @ 6 L/minute. Repeat investigations read Hb—9 g%, WBC—5,100, platelet—43,000 s/o thrombocytopenia. T. bilirubin—15 which was grossly elevated, SGOT/SGPT—46/44, S. creatinine—0.5 which were normal. The patient was posted for splenectomy in view of splenic infarct secondary to SCA. Post-splenectomy on day 5 patient developed bowel obstruction and thrombus formation in the infrarenal part of inferior vena cava (IVC). The patient was started on low molecular weight heparin (LMWH) injectable once daily. The patient was posted for emergency surgery in view of obstruction. Intra-op findings were s/o a small bowel band that was removed during the surgery. She was kept in the ward for further 4 weeks for postoperative management and monitoring. Her investigations on discharge were Hb—9 g%, WBC—7,000, platelet—118,000. T. bilirubin—1.2, SGOT/PT—37/35 which were normal. The patient was discharged after 14 days on tab hydroxyurea and other antibiotics.

## Discussion

The prevalence of sickle cell carriers among different tribal groups of India varies from 1 to 40%.[5] Madhya Pradesh has the highest load.[6] The patients here are prone to all the maternal complications like sickling crisis (both hemolytic and thrombotic) as well as infections. Our patient did not give any history suggestive of thrombotic problems but she had repeated attacks of jaundice which suggests repeated hemolytic episodes in the past. There is a higher maternal and fetal morbidity and mortality in this condition.[7] The mother's sickle cell percentage levels and hemoglobin levels were carefully monitored. To reduce hemolytic and thrombotic episodes, the modern management is to do exchange transfusions to keep the sickle cell percentage level below 20% and the hemoglobin level between 10 and 11 g/dL.[8] Although this patient did not develop erythrocytic antibodies due to repeated blood transfusions, it has been known to occur in Negro patients after repeated transfusions. This can lead to serious problems. Also, multiple blood transfusions and ongoing hemolysis due to crisis lead to elevation of serum bilirubin as evident in this case. Spontaneous abortion and intrauterine fetal demise are not that common in sickle cell carriers but in patients with SCD and sickle cell crisis, it has been reported previously too.[9]

As evident from the above report, we believe that pregnant women with SCD should be managed by a multidisciplinary team including hematologists and obstetricians with experience in high-risk pregnancy. The optimal approach of managing pregnancy complicated by SCD, which results in improved maternal and fetal outcomes, is yet to be identified. Some experts have tried various interventions, including prophylactic blood transfusion during pregnancy, but no difference was seen in pregnancy outcomes compared with women who were transfused on an episodic basis.[10–12]

The higher rate of fetal and maternal complications in pregnant women with SCD suggests a possible biological effect. A review by Rogers and Molokie suggested that pregnant women with SCD may face significant challenges from the normal physiological changes of pregnancy as a result of underlying end-organ damage initiated by SCD.[3] These changes significantly affect the cardiovascular, respiratory, and renal systems. Efforts to minimize these adverse events must, therefore, target these underlying biological factors that affect maternal and perinatal outcomes in women with SCD.

## Conclusion

This report highlights the overall increase in maternal and fetal complications associated with pregnancy in women with SCD. The higher rate of complications that occurs in women with sickle cell crisis exaggerated by underlying factors such as long-term anemia and pregnancy increases the danger further. Also, operative procedure in such patient results in further complications which may lead to long-term morbidity. Thus, a multidisciplinary approach with regular follow-up of SCD patients since the time of preconceptional time is vital to avoid pregnancy-related complications and also for a better pregnancy outcome.

## Clinical Significance

Sickle cell disease though rare can become a nightmare in long-term. The physiological changes of pregnancy like increased blood volume, increased metabolic demand, increased blood viscosity, and hypercoagulability get aggravated in SCD patients leading to increased incidence of complications. What we learned from our case is that multidisciplinary management is the key to managing such patients. Also, prepregnancy anemia and complications of a mother can further affect the outcome, thus preconceptional counseling is a crucial part of management.

## Acknowledgments

We would like to acknowledge the medicine department and surgery department of our college for successfully managing the patient medically and surgically.

## References

1. Piel FB, Hay SI, Gupta S, et al. 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.
2. Koshy M. Sickle cell disease and pregnancy. Blood Rev 1995;9(3):157–164. DOI: 10.1016/0268-960X(95)90021-7.
3. Rogers DT, Molokie R. Sickle cell disease in pregnancy. Obstet Gynecol Clin N Am 2010;37(2):223–237. DOI: 10.1016/j.ogc.2010.02.015.
4. Powars DR, Sandhu M, Niland-Weiss J, et al. Pregnancy in sickle cell disease. Obstet Gynecol 1986;67(2):217–228. DOI: 10.1097/00006250-198602000-00012.
5. Bhatia HM, Rao VR. Genetic atlas of Indian Tribes. Bombay: Institute of Immunohaematology (ICMR); 1987.
6. Rao VR. Genetics and epidemiology of sickle cell anemia in India. Indian J Med Sci 1988;42:218–222.
7. Tuck S, Studd J. Pregnancy in sickle cell disease in the UK. Br J Obstet Gynaecol 1983;90(2):112–117. DOI: 10.1111/j.1471-0528.1983.tb08893.x.
8. Tuck S, White JM. Sickle cell disease. Studd J, ed. Progressin Obstetrics and Gynaecology 1. Edinburgh: Churchill Livingstone; 1981. pp. 70–78.
9. Serjeant GR, Loy LL, Crowther M, et al. Outcome of pregnancy in homozygous sickle cell disease. Obstet Gynecol 2004;103(6):1278–1285. DOI: 10.1097/01.AOG.0000127433.23611.54.
10. Moussaoui DR, Chouhou L, Guelzim K, et al. Severe sickle cell disease and pregnancy. Systematic prophylactic transfusions in 16 cases. Med Trop (Mars) 2002;62(6):603–606.
11. Driss F, Tertian G, Becquemont L, et al. Management of high risk pregnancy in sickle cell disease by a strategy of prophylactic red cell transfusion or automated red cell exchange. Transfus Clin Biol 2007;14(4):386–392. DOI: 10.1016/j.tracli.2007.10.002.
12. Okusanya BO, Oladapo OT. Prophylactic versus selective blood transfusion for sickle cell disease in pregnancy. Cochrane Database Syst Rev 2016;12(12):CD010378. DOI: 10.1002/14651858.CD010378.pub3.