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Sickle cell disease in pregnancy and its outcome: An observational and prospective study conducted in tertiary care centre in central India

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Abstract
Background: Sickle cell anemia is autosomal recessive single gene defect in the ß-globin chain of HbA. Women with sickle cell disease are at greater risk of morbidity and mortality in pregnancy.
Objectives: To study the effect of Sickle cell Disease over the course of pregnancy, its outcome and management.
Method: It is a Hospital based observational prospective study conducted at Obstetrics and Gynaecology department Government medical college, Nagpur over a period of 18 months in 80 pregnant females having SCD attending OPD or admitted.
Result: An observational prospective study in pregnant females having SCD showed 22.5% Anaemia cases, 12.5% preeclampsia and cases for PROM was 2%. The Pre-term labour cases were seen only 5% and IUGR cases of 7%. The Birth Weight with <1.5kg was 5%, birth weight between 1.5-2.5 kg were 50% and birth weight greater than 2.5kg were 45%. The NICU Admission was 30%. Past history of crisis was seen in 15% of cases. Vaso-Occulusive Crisis was 6.3% and the hemolytic crisis was 6.3%. The Acute Chest Syndrome was 2.5%.
Conclusion: Comprehensive care may promote awareness of Sickle Cell Disease among affected women to present early for booking, assessment and management of symptoms.

Keywords: Sickle cell disease, complications, management, outcome

Introduction

Sickle cell disease (SCD) is an autosomal recessive disorder leading to the structural defect in the hemoglobin. It is the most common inherited hemoglobinopathy [1]. Sickle cell disease is homozygous HbSS. During pregnancy, it results in increased metabolic demands, hypercoagulable state and increased vascular stasis. Vasoocclusive crisis is more common in the later half of pregnancy. Studies on SCD pregnancy have focused on the risks including preterm labour, intrauterine growth retardation (IUGR), anaemia, gestational hypertension, pre-eclampsia, severe crises, postpartum hemorrhage, pulmonary diseases and infections, pyelonephritis, pulmonary infarction, pneumonia, acute chest syndrome, as well as fetal complications like preterm delivery, low birth weight, fetal distress in labour, increased perinatal mortality, pulmonary oedema.

Epidemiology: Highest prevalence among people in sub-Saharan Africa, South America, Central America, Mediterranean (Italian, Sicilian, and Greek), Middle Eastern, East Indian, Caribbean, and Central or South American descent. [2]Globally, India accounts for 14.5% of the total newborns with SCD.

SCD is prevalent in the tribal population of Odisha, Gujarat, Madhya Pradesh, Chhattisgarh and Rajasthan [3, 4]. It is estimated that 300,000 children are born each year with sickle cell disease (SCD), with 75% of them living in sub-Saharan Africa [5]. Prevalence of sickle gene is found to be 0-18% in north eastern India, 0-33.5% in western India, 22.5-44.4% in central India and 1-40% in southern India and the gene frequency of Hb-S varies between 0.031- 0.41 [6].

Pathophysiology: Sickle cell anemia is a disease of the erythrocyte, a genetic disorder caused by an autosomal recessive single gene defect in the ß-globin chain of HbA, which produces HbS. HbS is formed by the substitution of valine for glutamic acid in position six of the ß-globin chain of hemoglobin. The mutant ß hemoglobin subunits are normal in their ability to bind oxygen, less soluble in deoxygenated blood than normal hemoglobin., thus conditions of low oxygen tension results in the formation of intracellular, rod-shaped polymers. Repetitive cycles of sickling and polymerization however lead to membrane rigidity, and irreversible sickle cells are eventually formed. These permanently damaged erythrocytes are then cleared by the reticuloendothelial system. Thus, the average lifespan of the red blood cells of sickle cell patients is 17 days compared with the 120- day lifespan of normal erythrocytes. This results in a chronic haemolytic anaemia (haematocrit usually 20-30%) as the marrow's capacity to generate new red blood cells is limited.

Events during pregnancy

The steady state hemoglobin level falls during pregnancy reaching the lowest levels between 32 and 34 weeks, causing acute painful crisis which occurs secondary to vaso-occlusion and precipitated by infection, stress, dehydration and cold damp conditions. Acute chest syndrome is a sudden onset of pleuritic chest pain and dyspnoea that mimics pneumonia or pulmonary embolism precipitated by infection, marrow emboli, thromboembolism, or atelectasis. The combination of albuminuria and systolic hypertension occurring during a bone-pain crisis and should not be misdiagnosed as pre-eclampsia. Eclampsia was reported in 1 per cent and pre-eclampsia in 14 per cent of pregnancies. Many etiological factors causing preeclampsia occurs in SCD and this might explain why they are more likely to develop pre-eclampsia or have an exacerbation of pre-eclampsia. Bony abnormalities such as osteonecrosis of the femoral and humeral heads, renal medullary damage, autosplenectomy, (and splenomegaly in other variants), hepatomegaly, ventricular hypertrophy, pulmonary infarction, pulmonary hypertension, cerebrovascular accidents, leg ulcers and an increased predisposition to infection and sepsis, especially due to encapsulated organisms such as Streptococcus pneumoniae and Hemophilus influenzae can occur [9]. Blood transfusion is often necessitated in a number of HbSS patients.

Occasionally exchange blood transfusion may become necessary in those with severe vaso-occlusive crises and acute chest syndrome. The reason for exchange transfusion is to decrease the concentration of HbS, thus increasing the overall oxygen-carrying capacity of the blood, which will reduce the chances of sickling and therefore tissue damage, without increasing viscosity. Exchange transfusion is usually preferred in the acutely unwell patient with the sickle cell syndromes.

Most deliveries should be by the vaginal route. [7, 8] Caesarean section has been advocated for obstetric indications, disproportion due to pelvic outlet deformity and is increasingly used in an attempt to decrease the high fetal wastage in deliveries and in complications. Deliveries have also been reported to be complicated by retained placenta and still birth.

Diagnosis

The sickling test is a solubility test used in screening for sickle cell disease. Haemoglobin Electrophoresis (diagnostic test) is a method of determining the type and size of haemoglobin molecules in the blood, by observing the rates of transit of these negatively-charged proteins in an electric field medium. It is used to diagnose the haemoglobinothies. It is a blood test and requires a few millilitres of blood from a vein. DNA analysis provides the most accurate diagnosis in patients of any age, but it is still relatively expensive.

Genetic counseling

Two tests can be used to help expectant parents find out if their child is affected.

1. Amniocentesis, done usually at 14-16 weeks of pregnancy, tests a sample of the amniotic fluid in the womb. Test results often take 1- 2 weeks.
2. Chorionic villus sampling or CVS, involves the removal and testing of a very small sample of the placenta during early pregnancy. The sample, which contains the same DNA as the fetus, is removed by catheter or a fine needle inserted through the cervix or by a fine needle inserted through the abdomen. Results are usually available within 2 weeks.

Objectives

- To study the effect of SCD over the course of pregnancy
- To study the complication of SCD in antepartum, partum, post-partum and its management
- To Study effect of maternal SCD on Fetus and its outcome

AIM

- To improve the outcome of SCD in pregnancy
- To decrease the complications and its management during pregnancy.

Materials and Methods

- It is a Hospital based observational prospective study conducted at Obstetrics and Gynaecology department Government medical college, Nagpur during January 2020 to June 2021 in 80 pregnant females having SCD attending OPD or admitted over a period of 18 months.

Inclusion criteria

- All Pregnant women attending ANC OPD having SCD diagnosed by Hb-electrophoresis.
- Who are willing to participate in the study.

Exclusion criteria

- Women not willing
- Pregnancies which terminated in abortion

Observations and Results

Observational prospective study was conducted at Obstetrics and Gynaecology department Government medical college, Nagpur over a period of 18 months in 80 pregnant females having SCD attending OPD or admitted.

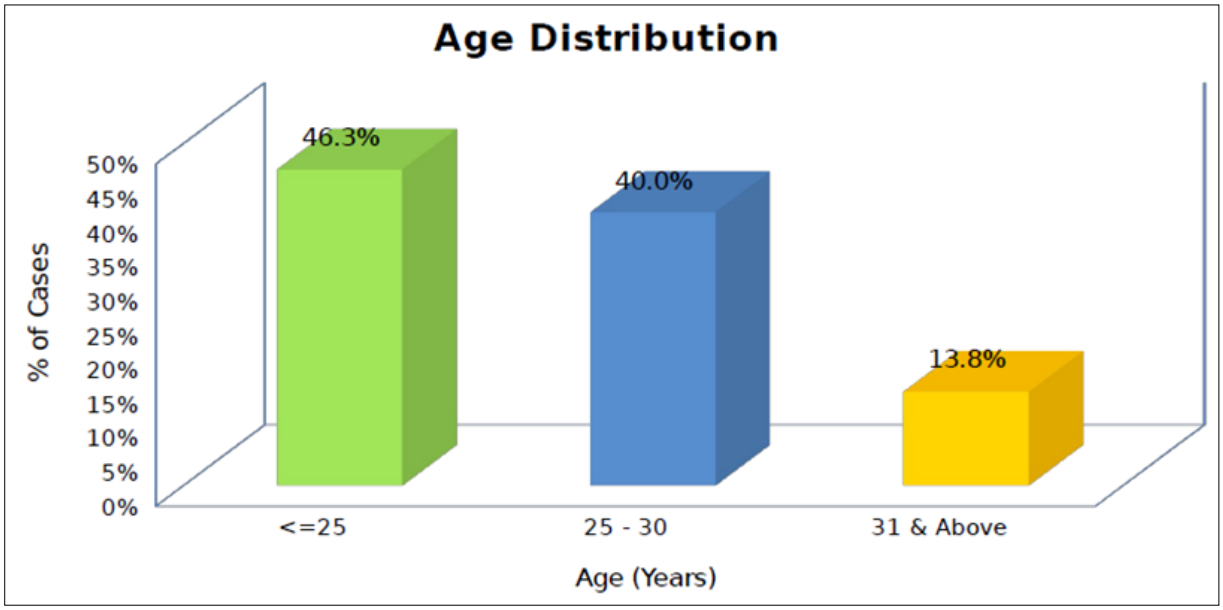

Fig 1: Age distribution

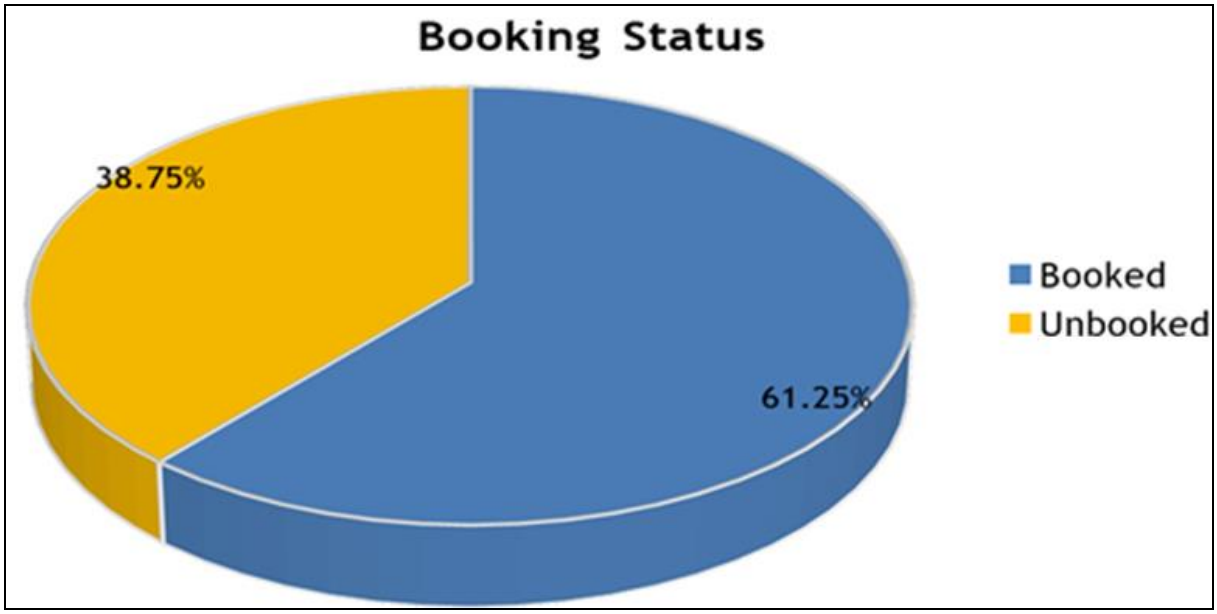

Fig 2: Distribution of study population according to booking status

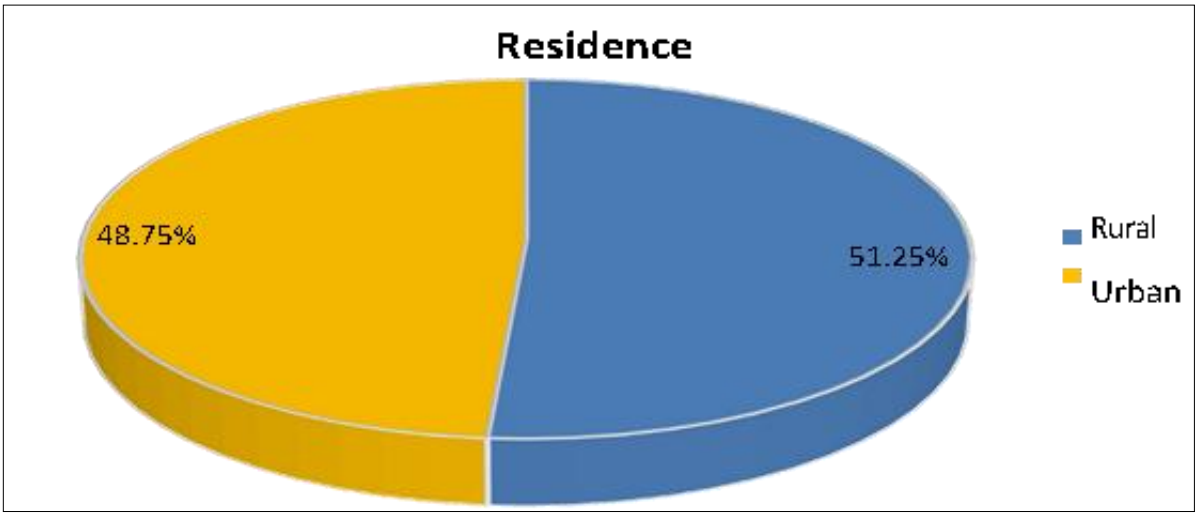

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Fig 3: Distribution of cases according to Residence

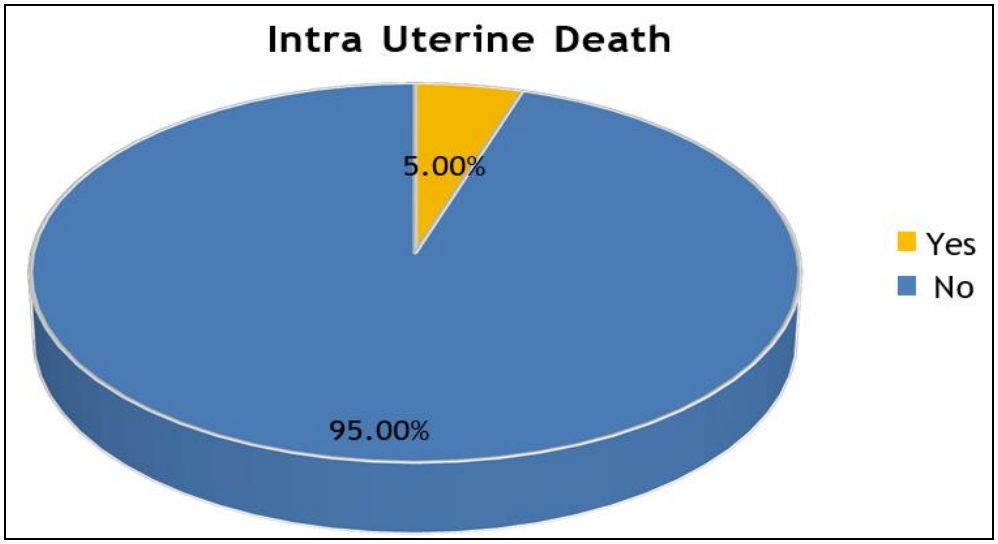

Fig 4: Distribution according to Intra Uterine Death

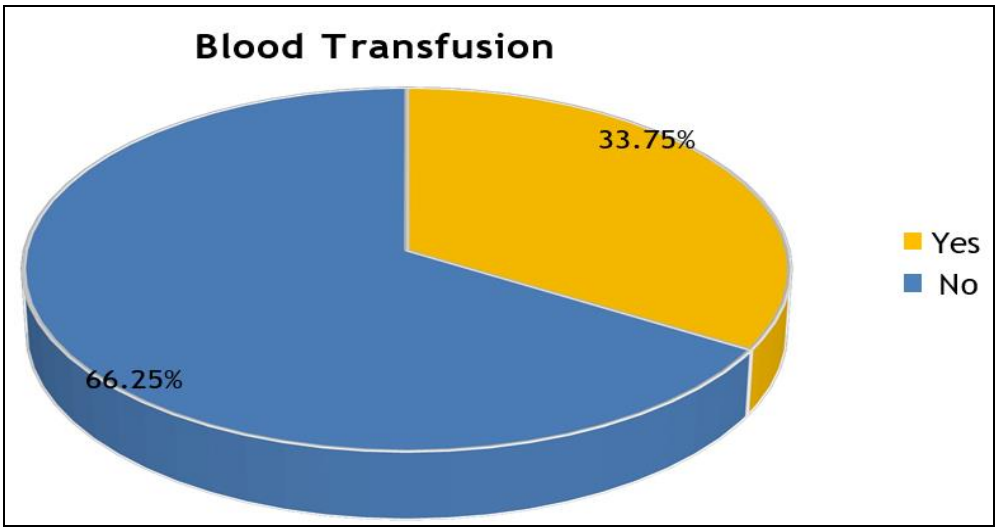

Fig 5: Distribution of cases according to Blood Transfusion

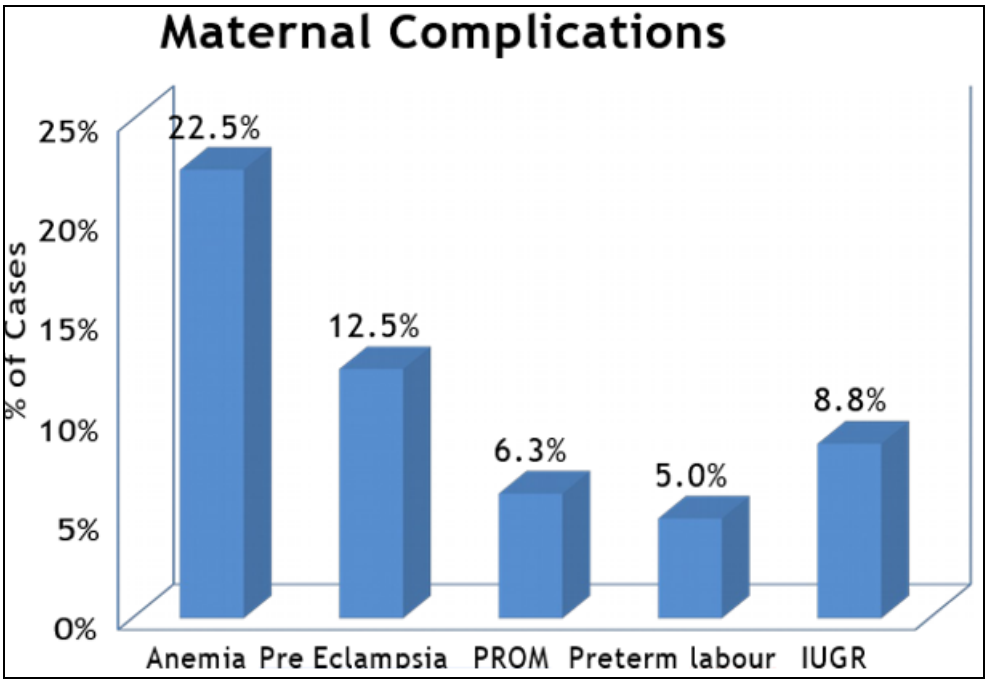

Fig 6: Distribution according to maternal complications

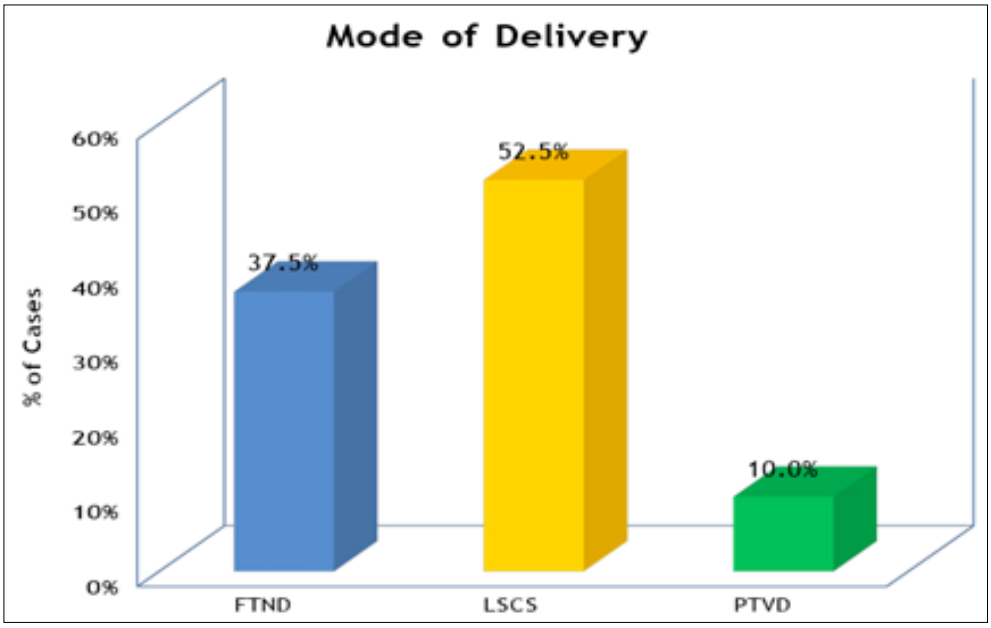

Fig 7: Distribution according to Mode of Delivery

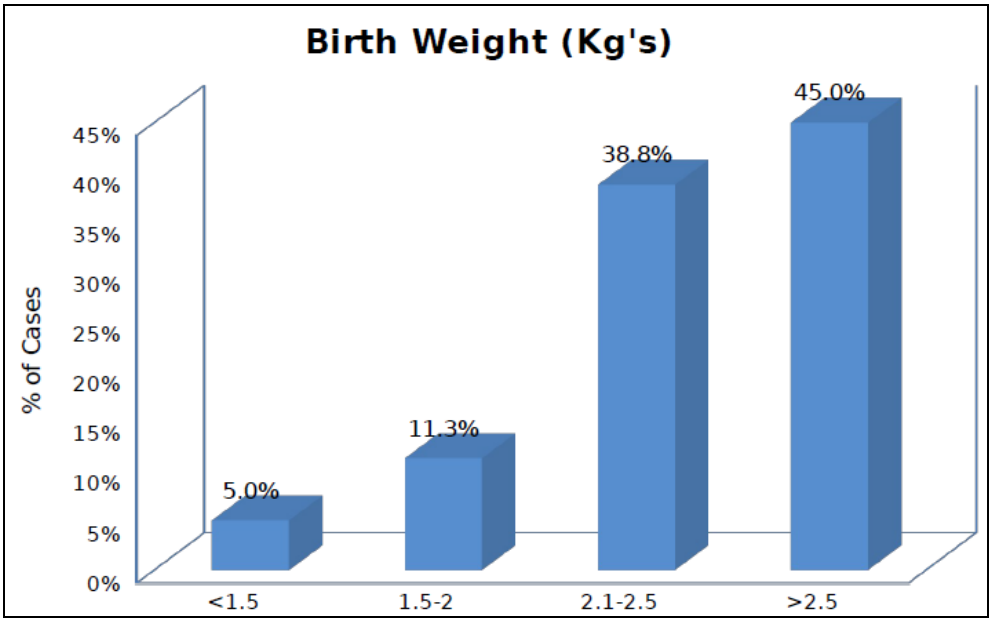

Fig 8: Distribution according to Birth weights

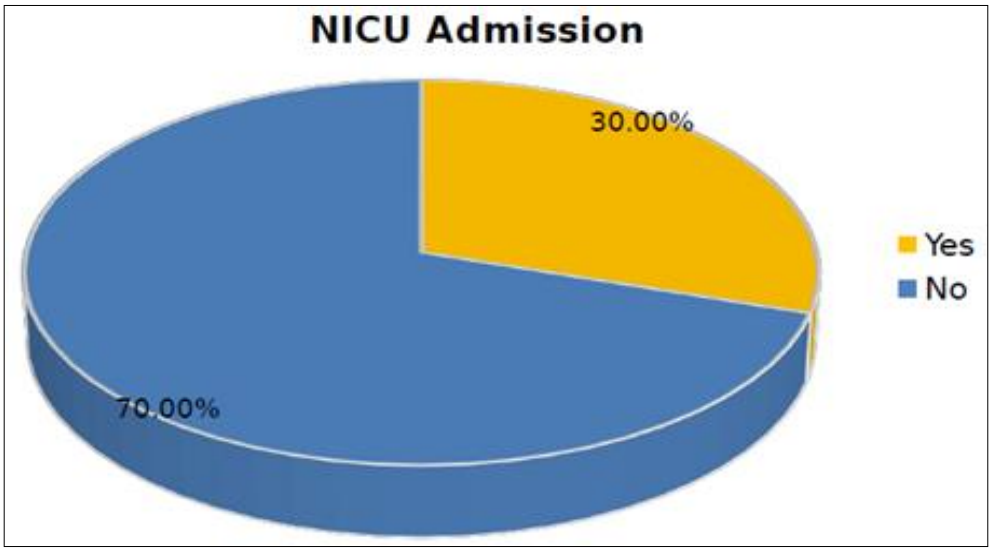

Fig 9: Distribution according to NICU Admission

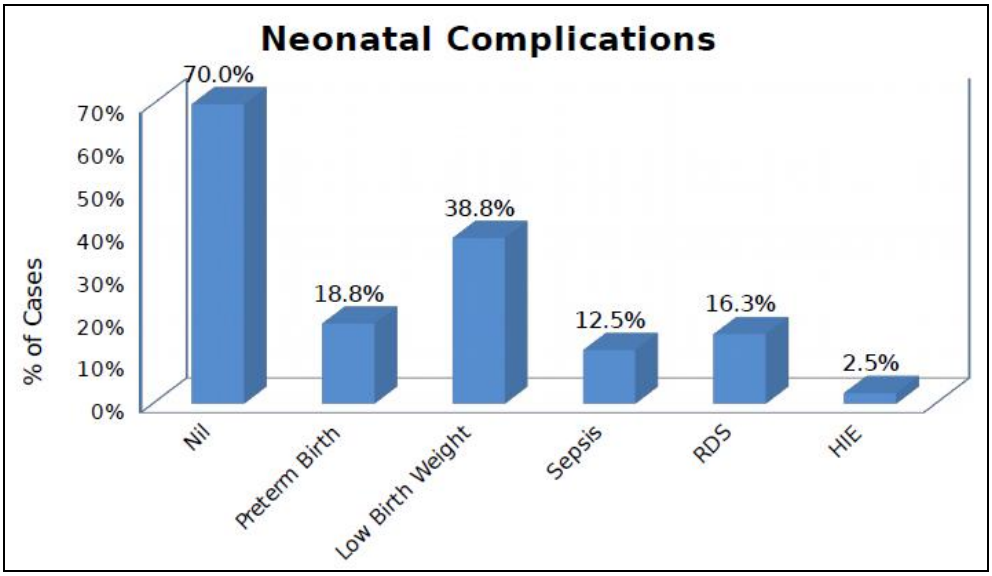

Fig 10: Distribution according to Neonatal complication

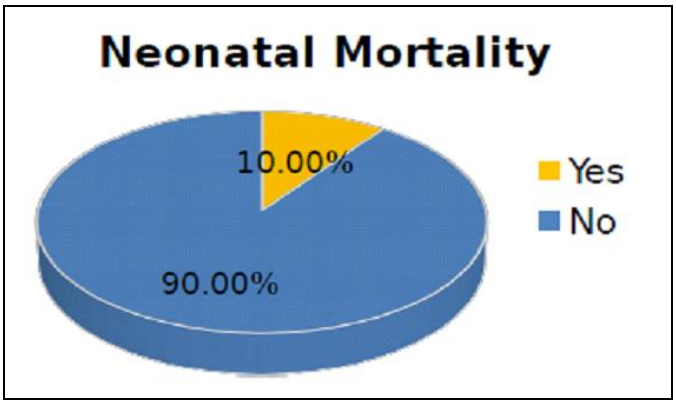

Fig 11: Distribution according to Neonatal Mortality

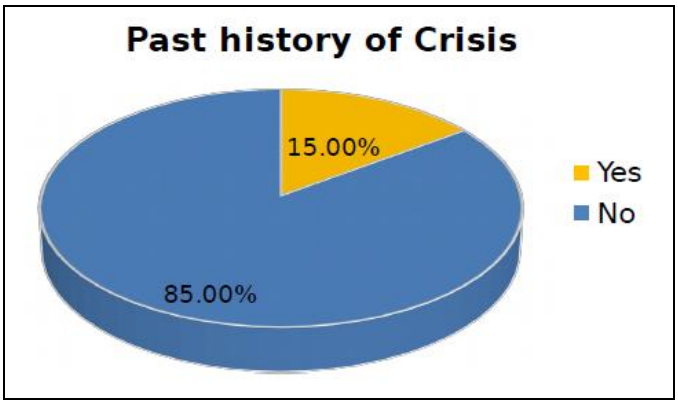

Fig 12: Distribution according to Past history of crisis

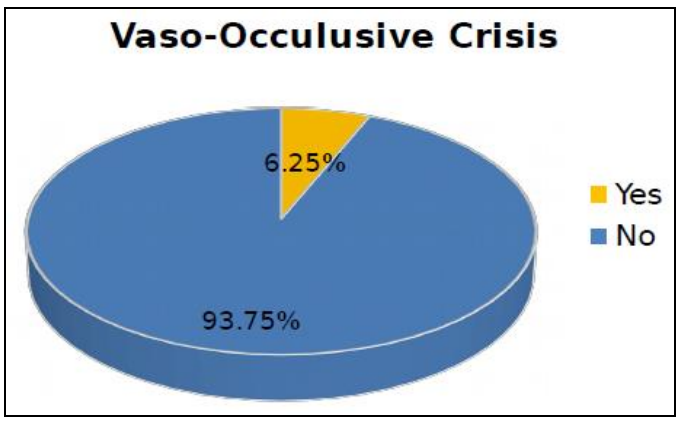

Fig 13: Distribution according to Vaso-Occulusive Crisis

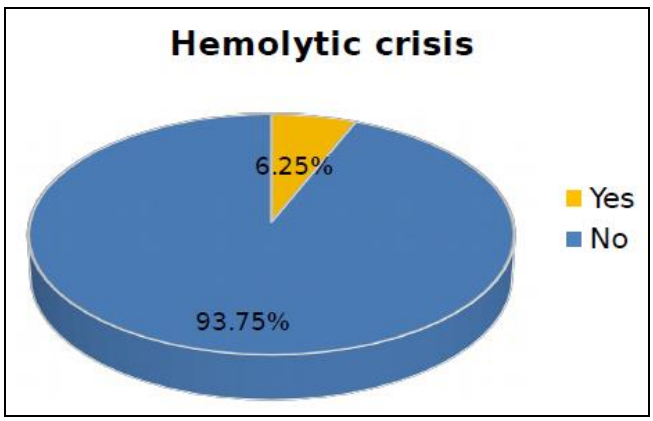

Fig 14: Distribution according to Hemolytic crisis

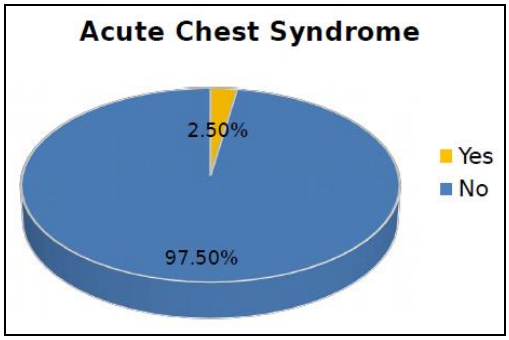

Fig 15: Distribution according to Acute Chest Syndrome

Discussion

An observational prospective study performed in tertiary care centre in pregnant women having SCD attending antenatal clinics. We analyzed the results of our studies and compared with a prospective study -pregnancy complications and outcomes in haemoglobin SS women conducted by DR. Monsurat Bolanle Aderolu in Lagos University at Nigeria [9].

It was found in this study that complication rate is higher in the HbSS pregnant women compared to their HbAA counterpart. In our study, patients who require blood transfusion is 33.8% while Dr. Monsurat Bolanle Aderolu found 26% in his study. 52.5% patient underwent lower segment cesarian section, 37.5%

underwent full term normal delivery and 10% underwent preterm vaginal delivery. While Dr. Monsurat Bolanle Aderolu found in his study that LSCS -68% and 20% vaginal delivery. We found in our study 5% intrauterine fetal demise whereas in Dr. Monsurat Bolanle Aderolu it is found to be 2%.

Table 1: Comparison of Mode of Delivery and Blood transfusion

	Blood Transfusion	LSCS	FTND	IVFD
Our study	33.8%	52.5%	37.5%	5%
Dr. Monsurat	26%	68.0%	20%	2%

Maternal complications: In Dr. Monsurat Bolanle Aderolu study the Anaemia was seen in 2% cases whereas in our study it is 22.5%. Also, our study shows 12.5% preclampsia cases but Dr. Monsurat Bolanle Aderolu 8% cases. The cases for PROM by Dr. Monsurat Bolanle Aderolu study were 6.3% whereas our study show 2%. The Pre-term labour cases are seen only 5% by our study but Dr. Monsurat Bolanle Aderolu study shows 28% of the cases. Our study shows IUGR cases of 7% but Dr. Monsurat Bolanle Aderolu study shows 16%.

Table 2: Comparison of Maternal complications

	Our study	Dr. Monsurat Bolanle Aderolu
Anaemia	22.5	2%
Preeclampsia	12.5%	8%
PROM	6.3%	2%
Pre-term labour	5%	28%
IUGR	7%	16%

Birth Weight Study: In Dr. Monsurat Bolanle Aderolu study, the Birth Weight <1.5kg was seen in 6% cases whereas in our study it is 5%. 32% cases in Dr. Monsurat Bolanle Aderolu have birth weight between 1.5-2.5 and 62% cases have birth weight greater than 2.5 kg. But in our study cases with birth weight between 1.5-2.5 are 50% and birth weight greater than 2.5 are 45%.

Table 3: Comparison of Birth weight

Birth Weight (Kg's)	Dr. Monsurat Bolanle Aderolu	Our study
<1.5	6%	5.0%
1.5-2.5	32%	50.0%
>2.5	62%	45.0%

NICU Admission: In Dr. Monsurat Bolanle Aderolu study the NICU Admission was seen in 42% cases whereas in our study it is 30%.

Table 4: Neonates with NICU Admission

NICU Admission	Dr. Monsurat Bolanle Aderolu	Our study
Yes	42.0%	30.0%
No	58.0%	70.0%

Past history of Crisis: We have observed that the past history of crisis was seen in 15% of cases.

Table 5: Cases with Past history of Crisis

Past history of Crisis	No. of Subjects	Percentage
Yes	12	15.0%
No	68	85.0%
Total	80	

Vaso-Occulusive Crisis: In Dr. Monsurat Bolanle Aderolu study the vasoocclusive crisis was seen in 32% cases whereas in our study it is 6.3%.

Table 6: Occurrence of Vaso Occlusive crisis

Vaso-Occulusive Crisis	Dr. Monsurat Bolanle Aderolu	Our study
Yes	32.0%	6.3%

Hemolytic crisis: In Dr. Monsurat Bolanle Aderolu study the hemolytic crisis was seen in 2% cases whereas in our study it is 6.3%.

Table 7: Occurrence of Hemolytic crisis

Hemolytic crisis	Dr. Monsurat Bolanle Aderolu	Our study
Yes	2%	6.3%

Acute Chest Syndrome: In Dr. Monsurat Bolanle Aderolu study the Acute Chest Syndrome was seen in 6% cases whereas in our study it is 2.5%.

Table 8: Occurrence of Acute chest syndrome

Acute Chest Syndrome	Dr. Monsurat Bolanle Aderolu	Percentage
Yes	6%	2.5%

We observed that maternal Anaemia preeclampsia and IUGR were more common maternal complications.

Unlike several studies that have reported increased incidence of spontaneous miscarriages, still birth and ectopic pregnancy in HbSS pregnant women, our study did not record any of these complications [5, 10, 11]. This is probably because the women studied booked after the first trimester above 16 weeks, when the risk of spontaneous miscarriages and ectopic pregnancies are less. This study also found that the HbSS parturients are more likely to have stable haematocrit post-partum probably because of the low threshold for blood transfusion in this category of women after delivery even before they become haemodynamically unstable. This practice is reflected in this study by the higher incidence of blood transfusion in the HbSS women 33.8%.

This study did not report any maternal death unlike previous studies [5, 6, 12, 13]. Probably because of the selection criteria which excluded high risk pregnant women with other co-morbidites except sickle cell disease. More so, the participants recruited were all booked and managed in tertiary institutions where they have access to multidisciplinary care.

5% of the cases shows intrauterine death. Low birth weight is one of the most consistent findings in neonates born to mothers with sickle cell disease 7,59,60. Almost half of the neonates in this study were low birth weight which reflected in the number of neonatal unit admissions. This study showed that HbSS pregnant women are more likely to have babies with birth asphyxia.

Conclusion

Although sickle cell disease poses higher obstetric risk in pregnancy, the maternal and perinatal outcome can be as good as in the non- sickle cell disease pregnant women if adequate and prompt health care is given to this group of women.

Present study showed anaemia and preeclampsia were found to be more commom maternal complications and thus early diagnosis is important for better outcome. Blood transfusion is important and beneficial. Hence, services at primary health care centres helps in early identification and referral of complicated cases to tertiary care centre.

Few cases showed IUGR, preterm and intra uterine demise. Half of the babies born to SCD mothers had birth weight between 1.5-2.5 kg. However, it is important to be vigilant at all times while managing HbSS pregnant women as labour complications

can arise anytime. Preconception care should be emphasized for this category of women (HbSS pregnant women) and multidisciplinary approach and prompt intervention are highly recommended to minimize perinatal and maternal deaths. Comprehensive care may promote awareness of Sickle Cell Disease among affected women to present early for booking, assessment, and management of symptoms. It is hoped that the results of this study will contribute to the existing knowledge about Sickle Cell Anaemia in pregnancy and help to strengthen the monitoring, management and follow up of this group of patients.

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