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A PROSPECTIVE OBSERVATIONAL STUDY OF INCIDENCE AND FETO-MATERNAL OUTCOMES OF SICKLE CELL DISEASE SPECTRUM IN PREGNANCY AT A TERTIARY CARE CENTRE OF CENTRAL INDIA

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ABSTRACT

Background: Sickle cell disease is a most important hemoglobinopathy encountered during pregnancy. The Sickle cell belt in Madhya Pradesh is characterized by a high prevalence of Sickle cell trait which occurs particularly in Betul, Chhindwara, Seoni, Mandala, Balaghat and Dindori District. All these patients are referred to our institute for for further management. **Objective:** This study aims to assess the incidence , complications and feto – maternal outcome of Sickle cell disease in pregnancy. **Method:** This is a prospective observational study carried out in our Department of Obstetrics and Gynecology, MTH Hospital and MGM Medical College, Indore from January 2022 to December 2022. This study is based on data collected during the course of their management and hospital stay. **Results:** In the prescribed period 452 antenatal and postnatal cases of Sickle cell disease were reported to our institute.Out of the total 10025 deliveries at our institute 452 cases were of sickle cell disease i.e – 4.0 %. As physiological anaemia is very common nearly 89% patients were anaemic and required blood transfusion. Majority of cases were belonging to rural population and were diagnosed sickle cell disease first time during pregnancy. Only 11% cases were known case of sickle cell disease. 16.89% cases of sickle cell trait and 83.11% cases were of sickle cell anaemia. Blood transfusion was done in 93% cases. 16.89% cases of sickle cell trait and 83.11% cases were of sickle cell anaemia. Blood transfusion was done in 93% cases.In terms of featl outcomes , 12.3% cases were pteterm still birth , whereas 15.7% cases were term stillbirth. 4.7% cases landed up into abortion. And 77.6% babies born were of low birth weight. **Conclusion:** Our study provides important insight into the challenges faced by women with sickle cell disease during pregnancy. We found that pregnant women with sickle cell disease had a higher risk of complications, such as preterm, birth, pre-eclampsia, fetal growth, restriction, still births as compare to women without the disease. The study also found that the majority of women with sickle cell disease do not receive appropriate care during pregnancy, highlighting the need for improved healthcare, infrastructure, timely and specialised care for this population.

KEYWORDS : Sickle cell disease, Pregnancy, Tribal Belt , feto- maternal outcomes.

INTRODUCTION

Sickle cell disease is the most important haemoglobinopathy encountered during pregnancy because of the severity of the complications associated with this condition. Pregnancy in women with sickle cell disease is associated with increase feto -maternal mortality and morbidity. Sickle cell disease is the most common inherited condition worldwide. It is a single gene disorder which is autosomal recessive inheritance. Sickle cell disease is a group of disorder which includes Sickle cell trait (HbSC) and Sickle cell anemia (HbSS) . Prevalence of sickle cell disease in Madhya Pradesh is nearly 10% .[1] The sickle cell belt in Madhya Pradesh is characterised by a high prevalence of sickle cell trade which occurs particularly in Betul, Chhindwara, Seoni, Mandala, Balaghat and Dindori districts. These patients are referred to our tertiary care centre for management.

The underlying defect in sickle cell disease is the decreased solubility of the sickle cell haemoglobin. Reduced oxygen environment can be caused by hypoxia , cool environment , acidosis and dehydration. Then the sickle cells become prone to break down that causes haemolytic anemia and often vasoocclusion in small blood vessels.[2] This causes most of the other clinical features include including acute painful crisis . It has been observed that sickle cell disease can significantly complicate normal pregnancy as it is a hypercoagulable state which exacerbates vasoocclusive crisis, stroke , acute chest syndrome , making them a high risk pregnancy. The later presents with, fever, tachypnoea , pleuritic chest pain , worsening anaemia and chest infiltrates and is the leading cause of maternal mortality. [3] Other complications include retinopathy, leg ulcers, cholelithiasis, stroke , pulmonary hypertension, renal papillary necrosis, avascular necrosis of femoral head, splenic, sequestration etc. Acute chest syndrome frequently complicates vasoocclusive crisis and multi Lobar involvement is common . Causes includes infection, infarction and fat embolism , which have a particularly poor prognosis. Acute sickling crisis occurs in almost 37% of pregnant women before delivery and 12% in the postnatal period.[4] Along with adverse maternal outcome , myriads of adverse fetal effects were also noted in terms of abortions, preterm labour, IUGR , preterm still birth, term still birth and increased SNCU admissions. [5]

MATERIAL AND METHOD

A prospective observational study was conducted in Department of Obstetrics & Gynaecology, M.Y. Hospital and M.T.H. Hospital, M.G.M. Medical College, Indore (M.P.)All the admitted women in Department of Obstetrics & Gynaecology, M.Y. Hospital and M.T.H. Hospital, Indore during the study period from January 2022 to December 2022 , formed our study population. Diagnosis of sickle cell disease was made on the basis of HB electrophoresis . Inclusion criteria included all pregnant and puerperal women having sickle cell disease as diagnosed by HB electrophoresis. A total of 452 cases of sickle cell disease were studied. All of these patients were investigated and managed accordingly. These patients were then followed up for complications and feto-maternal outcome. The relevant data was collected and classified on frequency distribution graph.

RESULTS AND OBSERVATION

Out of the total 10025 deliveries at our institute 452 cases were of sickle cell disease i.e – 4.0 % . As physiological anaemia is very common nearly 89% patients were anaemic and required blood transfusion. Majority of cases were belonging to rural population and were diagnosed sickle cell disease first time during pregnancy. Only 11% cases were known case of sickle cell disease.

Table 1

Status at admisssion	Frequency	Percentage
ANC	397	87%
PNC	16	13%

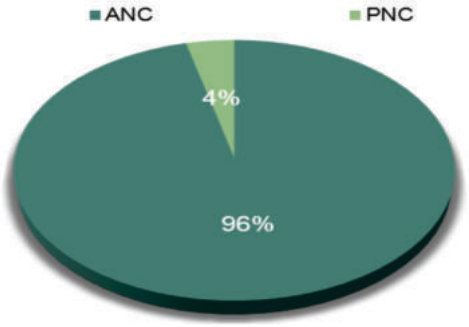


Table 2

Type	Frequency	Percentage	Mortality	Percentage
Sickle cell trait	321	71%	03	1%
Sickle cell disease	092	29%	04	12.5%

It has been observed that even sickle cell trait may have complicated pregnancy outcomes.

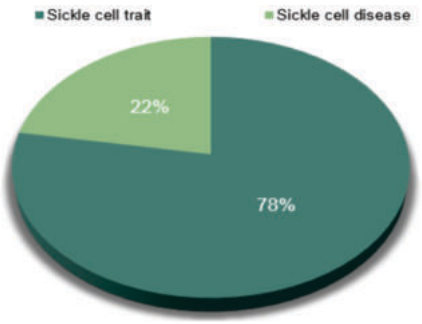


Table 3 Complications at the time of admission

Acute painful crisis	16	3.5%
Moderate and severe Anemia	347	76%
Acute chest crisis	13	2.5 %
Respiratory distress	18	4.5%
Avascular necrosis of femoral head	01	0.25%
Cortical necrosis of brain	01	0.25%
Renal papillary necrosis	02	05%
None	10	2.4%

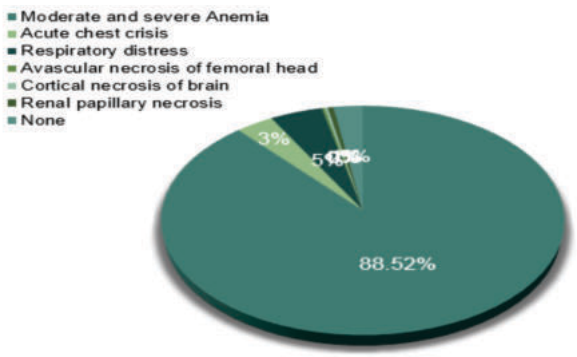


Table 4 Outcome of pregnancy

Outcome	Frequency	Percentage
Abortion	22	4.7%
Preterm still birth	56	12.3%
Term still birth	71	15.7%
Preterm live birth	109	24.1%
Term live birth	167	36.9%
Conservative management	27	5.9%

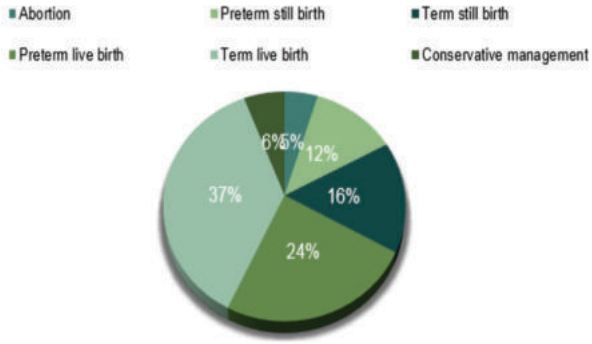
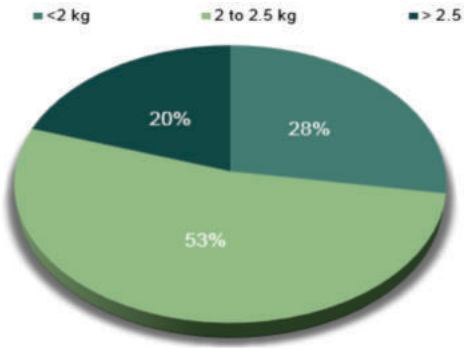


Table 5 Fetal outcome

Birth weight	Frequency	Percentage
<2 kg	111	27%
2 to 2.5 kg	212	52.60%
> 2.5	80	19.8



DISCUSSION

Sickle cell disease significantly complicated maternal and fetal outcomes in our studies. We have observed that 93% of the total cases that were reported to our Institute were referred from tribal belt of Madhya Pradesh 87% cases reported to us during their antenatal period where is 13% cases were post natal. Out of the total 10,025 deliveries 403 cases were of sickle cell disease which constitutes 4%. Out of all the 452 cases, 22% cases were of sickle cell trait, rest were Sickle cell anemia. This finding is consistent with the study conducted by G Desai et al. [6] Where 15.6% cases were of sickle cell trait. It is also similar to study conducted by S. Rajoria et al.[7] 16.89% cases of sickle cell trait and 83.11% cases were of sickle cell anaemia. Blood transfusion was done in 93% cases ,inspite of the fact that many of the patients were mildly anemic. Similar findings were observed by G Desai et al[6] where half of the deliveriers required Blood transfusion. 13% cases landed up into acute chest crisis whereas 16% cases landed up into acute painful crisis. S.Rajoriya et al [7]has also observed that 44.7% cases were associated with sickle-cell crisis. In terms of featl outcomes , 12.3% cases were pteterm still birth , whereas 15.7% cases were term stillbirth. 4.7% cases landed up into abortion. And 77.6% babies born were of low birth weight. All these findings were consistent with the study conducted by g.Desai et al where percentage of stillbirth is 9.9 where is 17.3% cases had preterm delivery and 70.2% cases were low birth weight . In study conducted by B.Subramania et al [8] They have also observed increased to risk of abortion, IUGR, still birth, adverse fetal outcome. D.Jain et al [2] has also observed

that 19% babies were still born.

CONCLUSION

1. Our study provides important insight into the challenges faced by women with sickle cell disease during pregnancy. We found that pregnant women with sickle cell disease had a higher risk of complications, such as preterm, birth, pre-eclampsia, fetal growth, restriction, still births as compare to women without the disease.
2. Most of the patients it belong to low socio-economic groups who were diagnosed or investigated for the first time during their pregnancy which shows the importance of regular antenatal check-ups that are still lacking in suck groups.
3. It has been observed that the prevalence of sickle cell disease is as high as 40% in certain tribal population of the state. The high prevalence is attribute it to combination of genetic and environmental factor, including consanguinity, malaria, endemicity and poor health infrastructure. Pregnancy adds up to the risk.
4. Lack of awareness and prenatal diagnosis made this population rich in clinically significant anemia from sickle cell trait . And thus, with advent of time, prevalence of sickle cell anemia has increased.
5. The study also found that the majority of women with sickle cell disease do not receive appropriate care during pregnancy, highlighting the need for improved healthcare, infrastructure, timely and specialised care for this population.
6. It is crucial to develop the effective intervention and strategies to address the unique needs of pregnant women with this disease. This may include developing specialise, clinics and programmes for antenatal care, providing education and awareness to health care providers as well as improving access to essential medication and treatment.
7. It is essential for women with sickle cell disease who are considering pregnancy to receive appropriate preconceptional counselling and care to optimise their health and reduce the risk associated with pregnancy.

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