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Study of perinatal outcome in pregnancy with sickle cell disease

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ABSTRACT

PURPOSE To study the perinatal outcomes and complications of pregnancy in babies born to mothers with sickle cell anaemia or trait and . To establish a comparison between this study and the previous studies done on this topic. **MATERIALS AND METHODS**-A Case control study conducted in Department of Obstetrics and Gynecology, Acharya Vinoba Bhave Rural Hospital(A.V.B.R.H.)wardha,India. **RESULT**-Maternal complication mainly found were, anaemia preeclampsia, and pregnancy-induced hypertension whereas growth restriction prematurity were found predominantly in fetuses of sickle cell positive patients. **CONCLUSION** Sickle cell hemoglobinopathy is a common disease in this part of central india(wardha district)with the prevalence being 5.7%.pregnancy with sickle cell disease is found to be associated with fetal/maternal complications.

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1. Introduction

Sickle cell disease in pregnancy still remains a major challenge for gynecologists, trying to improve the quality and duration of life of both the affected mother and the foetus. The world population report (1975) gives the incidence of anemia to be 100% among pregnant women in India⁵. Sickle cell haemoglobinopathy is a very common disease in this belt of central India (Wardha district) and is very prevalent (5.7%) in pregnant woman attending the ante natal OPD having either SS or AS character¹.

Sickle cell disease is an important hereditary haemoglobinopathy, a type of disease characterized by production of defective haemoglobins¹. Sickle cell hemoglobin is produced by substitution of valine by glutamic acid at position 6 of β chain of the

normal hemoglobin. Gene mutation – when homozygous, the individual has sickle cell anaemia (Hb SS); when heterozygous, the individual has sickle cell trait (Hb AS)². The abnormal HbS tends to polymerize on deoxygenation and red blood cell containing HbS becomes less pliable and consequently deforms into the sickle shape. Sickle cell disease is a multi system disorder and risk of sickle cell anaemia during pregnancy includes an increase in gestational hypertension, preterm birth and small-for-gestational-age infants, chronic hemolysis, postpartum hemorrhage, repeated infections, growth retardation in addition to an acute life threatening complication called Crisis. Pain from ischemic necrosis of bone

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marrow or other organs usually becomes more frequent. Pulmonary complications are also common. Risks of maternal mortality are increased. Fetal wastage is also common and more than one third of pregnancy in woman with sickle cell syndrome have terminated in abortion, still birth or early neonatal death³. Low birth weight babies were born to SS mothers due to premature deliveries and fetal growth retardation⁴. Perinatal mortality is also very high. Hence this study was undertaken to assess the complications arising in perinatal period, the pregnancy outcome in sickle cell disease women and ways and methods through which such complications can be avoided or minimized to decrease perinatal abnormalities .perinatal period, the pregnancy outcome in sickle cell disease women and ways and methods through which such complications can be avoided or minimized to decrease perinatal abnormalities perinatal period, the pregnancy outcome in sickle cell disease women and ways and methods through which such complications can be avoided or minimized to decrease perinatal abnormalities.

AIM AND OBJECTIVES

1. To study the perinatal outcomes and complications of pregnancy in babies born to mothers with sickle cell anaemia or trait
2. To establish a comparison between this study and the previous studies done on this topic

Materials and Methods

Study Setting: This is a study conducted in Department of Obstetrics and Gynecology, Acharya Vinoba Bhave Rural Hospital (A.V.B.R.H.), a tertiary teaching hospital of Datta Meghe Institute of Medical Sciences University and its peripheral subcentres In Study approvals; This study has been approved by the Institutional Ethics Committee (IEC) of Datta Meghe Institute of Medical Sciences University.

Study type: A Case control study. **Sample Size:**A Sample of 50 cases, which include cases and controls. **Duration of Study:**Two months of data collection.One month for study analysis.

Consent: tests and their involvement for study purposes, during study period had been taken. been assured. **Inclusion Criteria:** All the pregnant cases, admitted to the obstetric wards or labour room.

Exclusion Criteria:

1. Patients not having consented to the procedure.
2. Patients with any other associated medical condition or disease.

Study procedure

All the pregnant females admitted to the obstetrics ward of Acharya Vinobha Bhave Rural Hospital (AVBRH) and its peripheral sub centers, in the duration of 6 months from January to June 2009 were studied retrospectively. Also, those admitted in these centers in the months of July and August 2009 were studied prospectively. In the retrospective study, previous medical records and case files of the pregnant women were studied and assessed and all the sickling positive women were included as cases. Due consent for this was

taken from the Medical Record Department of Acharya Vinobha Bhave Rural Hospital. In the prospective study, written consent was taken from all the pregnant women on an appropriate consent form. All the pregnant women who consented were screened for sickle cell disease (AS & SS character) by the sickling test. Positive cases were included as cases. These were then subjected to Hb electrophoresis to differentiate sickle cell anemia (SS) from sickle cell trait (AS) The added sum of the positive cases of the prospective and retrospective study were considered as the total number of cases in the study. Equal number of controls, negative for sickling and comparable with cases, were randomly recruited in the study. Mothers were regularly screened for any developing complications. After delivery, the babies were examined on parameters like Fetal outcome (Live birth, still birth, or neonatal death.) Gestational age (Preterm or term), Birth weights, APGAR scoring Requirement for NICU admission, Any other congenital anomaly. Thus perinatal outcome of pregnant females with sickle cell disease (SS and AS) was evaluated. Collection, Presentation and Interpretation of Data The collected data has been depicted in tabular form and with the use of graphs interpreted statistically and analyzed.

Statistical Analysis:

No statistical method has been used to analyse the results, as the sample size is small.

Observations and results are presented in tabulated form.

Table I Incidence of sickling character as per electrophoresis

SICKLING	NO OF CASES	%
AS	21	84
SS	4	16
TOTAL	25	100
controls	25	100

Table 2. shows the mean hemoglobin levels in the SS (homozygous sickling) group (7.5 g/dL) was lower as compared to that in AS (heterozygous sickling) group (8g/dL) and to that in the AA (non-sickling controls) group (9.1g/dL). Similar results were given by Sonwane et al³, with mean Hb level in SS (7.65 g/dL) was lower than that in AS (8.77 g/dL) and to that in

Table II. Hemoglobin levels of pregnant mothers.

HB	SUBJECTS				CONTROL	
	SS		AS		AA	
gm/dl	NO	%	NO	%	NO	%
<8	3	75	15	71.42	3	12
8-10	1	25	5	23.82	18	72
>10	0	0	1	4.76	4	16
TOTAL	4	100	21	100	25	100

Table 3. shows the Complications arising during pregnancy:Among these complications, Crisis was found in 25 %cases in SS group. Crisis as complication during pregnancy is reported to be 40% by Sonwane et al3, 48.6% by Dare et al22,28% by Chhabra et al23, 88% by Leborgne et al12.Cases of urinary tract infection were more in SS and AS groups as compared to AA group. Prevalence of UTI was25% in SS and 33.3% in AS. Whereas it was only 1.07% inAS women as per Sonwane et al3. There were 25% cases of Pre-eclampsia in SS and14.3% in AS groups with no incidence of the same in thecontrols. According to Sonwane et a3, pre-eclampsia was more in AS group (36.55%). In women with sickle cell disease it wasobserved to be 2.4% by Idrissa et al27, 16.2% by Dare et al22 ,12.62% by Deshmukh et al 26. Severe anemia (Hb<7g/dL) was found in 75% in SS,28.6% in AS, and 4% in AA cases. These results showextremely high prevalence of the condition but prevalence of only 24% in SS group and 11.82% in AS group as reported bySonwane et al3 ,and only 32% in SS and 5.74% in AS as per Kale et al4.

Table III. Complications in mothers during pregnancy and child birth.

COMPLICATIONS	SUBJECTS				CONTROL	
	SS	AS	AS	AA	NO	NO
	NO	%	NO	NO	%	NO
CRISIS	1	25	0	0	0	0
URINARYTRACT INFECTION	1	25	5	33.3	3	12
SEVEREANAEMIA	3	75	6	28.6	1	4
PREECLAMPSIA	1	25	3	14.3	0	0

Table 4. shows that the choice of mode of delivery in sicklingmothers was Cesarean section, which was done in 100 % in SS group and 38.10% in AS Group and 6.24% in AA group. As per Kale Ashish et al4 36% of SS cases ,16.9% of AS cases, and16.07% of AA cases had cesarean section. 64% SS cases 46.24%AS cases according to Sonawane anju et al5.

Table IV-mode of delivery

DELIVERYMODE	SUBJECTS				CONTROL	
	SS	AS	AS	AA	NO	NO
	NO	%	NO	NO	%	NO
LSCS	4	100	8	38.1	6	24
VAGINALDELIVERY	0	0	12	57.14	18	72
INSTRUMENTAL	0	0	1	4.76	1	4
TOTAL	4	100	21	100	25	100

Table 5. shows the fetal outcome among SS and ASpregnancies and Intra uterine death was noticed in 25% and 4.76%cases respectively, and rest being live births. Whereas only 4% and 1.14% IUD's were noticed in SS and AS groups according to KaleAshish et al4. When compared to the control (AA) group

(5.2%)there were significantly more preterm deliveries in the SS group(50%) and the AS group (9.53%) .There were more preterm deliveries in the SS group (50%) than the AS group (9.53%).Inwomen with sickle cell disease, preterm deliveries are reported to be72% by Sonwane Anju et al3, 21.6% by Dare et al6,20% by Chhabra et al7, Howard et al24,and 21% by Leborgene et al5, 56%by Kale Ashish et al 4 in SS group.Incidence of low birth weight babies(LBW) (<2500gm) which was 100% in SS and AS group and 16%in the control group, justifying higher risk of LBW in SS and ASgroup than AA group. Similar findings were observed by Sonwane Anju et al3 with 77.78% LBW in SS group and 48.45% in AS group.According to Kale Ashish et al4 LBW in SS,AS, and AA groups are56%,34.48% and 23.21% respectively. Similarly 31% babies in study of Poddar et al 8. Among the SS group 2 (66.6%) cases have reported to have moderately low APGAR score (below7), whereasin AS group 5% cases have severely low (below 3) and 5% caseshave moderately low score.

Table V-fetal outcome

Fetal outcome	SUBJECTS				CONTROL	
	SS	AS	AS	AA	NO	NO
	NO	%	NO	NO	%	NO
Live birth	3	50	20	95.24%	25	100
preterm	2	50	2	9.53%	2	8
term	2	75	19	90.47%	23	92
Live birth	3	25	20	95.24%	25	100
Intrauterine fetal death	1	25	1	4.76%	0	0
Birth wt<1500	1	25	2	9.52	1	4
Birth w1500-1999	1	50	1	4.76	2	8
Birth w2000-2499	2	0	18	85.72	1	4
Birth w>2500	0	0	0	0	21	84
APGARSCORE 0-3	0	66.6	1	5	1	4
APGARSCORE 4-7	2	33.4	1	5	2	8
APGARSCORE 8-10	1		18	90	22	88

Table 6. gives the Neonatal complications which includesjaundice developing in 25% cases in SS and 4.76% cases in AS groups. According to Kale Ashish et al4 jaundice developed in 27.12% and 8.43% in SS and AS groups respectively.

Table VI. Neonatal complications

APGARSCORE	SUBJECTS				CONTROL	
	SS	AS	AS	AA	NO	NO
	NO	%	NO	NO	%	NO
Septecemia	0	0	0	0	1	4
Jaundice	1	25	1	4.76	10	4

CONCLUSION

Sickle cell hemoglobinopathy is a common disease in this part of central India (Wardha district) with the prevalence being 5.7%. Pregnancy with sickle cell disease is bound to be associated with fetal/maternal complications. Vaso-occlusive crisis, anemia and preeclampsia were main maternal complications in our study. Complications lead to more preterm and caesarian deliveries. IUGR, IUFD, prematurity, low APGAR scores were found as main fetal complications in our study, requiring more NICU admission of neonate. In our study perinatal outcome is better in sickle cell trait than in sickle cell disease mothers. Meticulous antenatal care, along with close hematologic consultation is strongly recommended, and has proved in our study to reduce maternal morbidity and mortality. Further research needed on a larger scale to improve the fetal/maternal outcome in this disease.

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