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Original Article

Perinatal outcome in pregnancy with sickle cell anemia

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Abstracts

Objectives: To determine the perinatal outcome in pregnancy with sickle cell anemia. *Methods:* This is a comparative case control study, for which 1412 of pregnant women were chosen from ANC OPD obstetric ward and labor room. All of them were screened for sickle cell anemia, as the prevalence rate in this part of Orissa is 8% of the pregnant women, 112 women were found to be positive for either sickle cell disease or trait, confirmed by electrophoresis. Equal number of controls were randomly recruited in the study and matched for age, gravidity and other demographic factors. Perinatal outcome as regards birth weight, Apgar score, NICU admission and perinatal loss was compared in both the groups. Wherever applicable, chi-square test was applied. *Results:* The overall incidence of sickle cell anemia was 7.93%. Out of 112, sickle cell disease (SS) was found in 25 cases (22.32%) and sickle cell trait (AS) in 87 cases (77.67%). Prematurity was detected in 44% of (SS), 26.42% of (AS) and 17.8% in control group. Low birth weight babies born to SS, AS and controls were 56%, 34.48%, and 23.21% respectively. Perinatal mortality was 20% in (SS), 8.04% in (AS) and 6.25% in controls. *Conclusions:* The incidence of preterm deliveries, perinatal mortality and low birth weight babies are significantly high in women with sickle cell disease compared to controls. However the perinatal outcome in mothers with sickle cell trait is better as compared to sickle cell disease.

Key words: sickle cell disease, sickle cell trait, prematurity, low birth weight babies.

Introduction

Sickle cell hemoglobinopathy is a common disease in western Orissa and it is found that about 8% of pregnant women attending antenatal OPD have either SS or AS. In view of the high prevalence of the disease in this part of the country, this study has been undertaken to observe perinatal outcome of pregnancy in mothers with sickle cell disease and trait and compare it with controls.

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Sickle cell hemoglobinopathy is one of the common autosomal recessive heritable diseases characterized by production of abnormal sickle hemoglobin (HbS). The abnormal HbS tends to polymerize on deoxygenation & red blood cells containing HbS become less pliable and consequently deform into characteristic sickle shape, after which the disease is named. A person who inherits an abnormal HbS gene from one parent becomes the less harmful carrier state of sickle cell trait (AS). Inheritance of abnormal gene from both parents results in homozygous state of sickle cell disease (SS).

Sickle cell disease is a multisystem disorder whose clinical manifestation includes chronic hemolysis, repeated infections, growth restriction in addition to an acute life threatening complication called crisis which

is associated with considerable morbidity and mortality. Fetal wastage in the form of abortion, stillbirth and neonatal death is very high in SS women and ranges from 27% to 45%. Low birth weight babies were born to SS mothers due to premature deliveries and fetal growth retardation.

Methods

This was a case controlled study, carried out in the Department of Obstetrics and Gynecology, Burla Medical College. Total of 1412, pregnant women attending the ANC OPD or admitted to obstetric wards or labor room were screened for sickle cell anemia. Cases that were positive for sickling test were subjected for Hb electrophoresis to differentiate SS from AS. Eighty seven mothers with AS and 25 mothers SS were identified. All these cases were subjected for detailed examination and investigations like urine examination to detect bile salts by Hay's test, bile pigment by Fouchet's test & urobilinogen by Ehrlich aldehyde test. Diagnostic hemoglobin estimation and total leucocytic counts were done to rule out infection. USG was done routinely in all cases for fetal growth retardation. 112 cases that matched for age, parity, socioeconomic status, negative for sickling test with no other obstetric risk factor were studied as controls.

After delivery, the babies were weighed. Maturity of newborn babies was assessed and Apgar scoring was done. Resuscitation was done in babies with low Apgar. Selected cases were referred to NICU of VSS Medical College, Burla. The newborn babies were followed for seven days after the delivery for any neonatal complication.

Table 1. Birth weight of babies born to mothers with SS, AS & Control.

Birth Weights	S S n=25		AS n=87		Control n=112	
	No	%	No	%	No	%
<999 gm	4	16	6	6.89	5	4.6
200-2499 gm	10	40	24	27.58	21	18.75
≥2500 gm	11	44	57	65.51	86	76.78
LBW<2500 gm	14	56	30	34.48	26	23.21

Table No. 2. Gestational age of babies born to mothers with SS, AS & Control.

Gestational age	S S n=25		AS n=87		Control n=112	
	No	%	No	%	No	%
Preterm	14	56	83	95.4	21	17.8
Term	11	44	4	4.6	81	82.2

Table 3. Fetal outcome.

Fetal Outcome	S S n=25		AS n=87		Control n=112	
	No	%	No	%	No	%
Live birth	22	88	83	95.4	109	97.32
IUD	01	4	01	1.14	01	0.89
Still birth	02	8	03	3.44	02	1.78
Early neonatal death	02	8	03	3.44	04	3.57

Table 4. Newborns requiring NICU.

Duration	SS n=25		AS n=87		Control n=112	
	No	%	No	%	No	%
<24 hrs	2	9.09	9	10.84	11	10.09
24 hrs - 7 days	10	45.45	17	20.48	13	11.92

Table 5. Neonatal complication.

Complications	SS n=25		AS n=87		Control n=112	
	No	%	No	%	No	%
Hypoxic ischemic encephalopathy	4	18.18	6	7.22	4	3.66
Neonatal septicemia	2	9.09	3	3.61	2	1.83
Jaundice	5	22.72	7	8.49	2	1.83
Anemia	3	13.63	2	2.4	1	0.91

Results and Discussion

Out of total 1412 cases, the incidence of sickle gene in the present study was found in 112 cases (7.93%) of which sickle cell disease (SS) was 25 cases (22.32%) & sickle cell trait (AS) was 87 cases (77.67%). The mean age of pregnant mothers with SS, AS & controls were 24.2, 24.11 and 24.09 years respectively. The mean gravida of SS, AS & control group of women were 1.84, 1.98 and 2.06 respectively. The mean parity of SS, AS and control mothers were 0.68, 0.87 and 1.32 respectively.

In the SS group, 32%, in AS group 5.74% & in control group 3.57% of women were severely anemic. Cesarean section was done in 36%, 16.09%, 16.07% cases of SS, AS & control group respectively. Instrumental deliveries were 12%, 4.59% and 6% in SS, AS and control group respectively.

Table No.1 shows the birth weight of babies among the groups. Low birth weight babies born to SS, AS and control group mothers were 56% 34.48% and 23.21% respectively. Low birth weight babies are significantly more ($P<0.05$) in SS group. There was no significant statistical difference between the AS and control groups.

Sonawane [1] reported that 77.78% of low birth weight babies were born to SS mothers and 48.45% in the AS group. Similarly 31% babies in study of Poddar et al [8]. High incidence of LBW babies was due to fetal hypoxia throughout the pregnancy caused by anemia and fetoplacental insufficiency due to degree of sickling and vasocclusion in placental circulation.

From Table No. 2 it is seen that prematurity was detected in 44% cases of SS group ($P<0.05$), 26.42% cases of AS group and 17.08% cases of control group.

Preterm deliveries were more frequent in SS i.e. 44%, than 26.42% in AS and 17.8% in control. In women with SS, preterm deliveries are reported to be 21.6% by Dare et al [3], 20% by Chhabra [4], 23% by Howard[5], 21% by Leborgne-Samuel et al [6] and 72% by Sonawane [1].

Total perinatal mortality as seen in Table No. 3 was 20% in SS, 8.04% in AS and 6.25% in control groups. Early fresh still births were 8%, 3.44% and 1.78% in SS, AS and control groups respectively, suggesting intrapartum hypoxia. Fetal heart abnormalities developed in 27.27% cases of SS, 22.04% of AS and 12.84% in control group.

Table Nos. 4 and 5 show requirement of NICU and

neonatal complications respectively. Out of 25 new borns of SS mothers, 10(45.45%) babies required specialized care for more than 24 hours whereas 17(20.84%) babies of AS group and 13(11.92%) babies of control group required NICU for more than 24 hours.

Total perinatal mortality in the present study is 20% in SS group and 8.04% in AS group with that of 17.1% in SS group as reported by Poddar et al[2].

Table No. 5 shows that hypoxic ischemic encephalopathy was seen in 18.18% cases of SS group born babies, 7.22% and 3.66% babies of AS and control group respectively. Jaundice developed in 27.12%, 8.43%, 1.38% babies born in SS, AS and control group respectively. Similarly anemia was seen more in SS mothers.

In the SS group, 32% women were severely anemic, whereas in AS and control group 5.74% and 3.57% were severely anemic respectively. This is comparable to the Hb level in the study conducted by Sonawane[1]. The various factors that contribute towards the causation of anemia in these women are continued hemolysis, sequestration crisis, folate deficiency and iron deficiency. In the present study, the anemia was mostly normochromic because majority of the SS women were on iron and folate supplementation.

Cesarean section was done in 36%, 16.9%, 16.07% cases of SS, AS and control group respectively. Cesarean section was required in 16 out of 25(64%) in the SS group, and 43 out of 93 (46.24%) in the AS group in Sonawane's study. The cesarean section rate is reported to be 14.6% by Idrisa[7], 29.7% by Dare[3], 12% by El Shafei[8], 66.66% by Howard[5], 10.48% by Leborgne-Samuel et al[6], and 43.2% by Odum et al[9]. 24% cesarean sections were done in SS group for fetal distress as compared to Dare[3] it was 18.1%, 67% in El Shafei[8].

Conclusion

Sickle cell hemoglobinopathy is a common disease in the pregnant women of western Orissa. The incidence of sickle cell disease and sickle cell trait was found to be 1.77% and 6.16% in these pregnant women respectively.

The incidence of preterm deliveries, perinatal mortality and low birth weight babies and incidence of cesarean section in women with sickle cell disease is significantly high compared to controls. However the perinatal outcome in mothers with sickle cell trait is favorable compared to sickle cell disease cases.

Screening of all cases of anemia in antenatal cases in prevalent area and early, aggressive and comprehensive perinatal care can improve the perinatal outcome in these high risk mothers.

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