Outcome of Pregnancy in Sickle Cell Trait and disease in Western Orissa.

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Summary: This prospective study was conducted in the Department of Obstetrics & Gynaecology and in the sickle cell Research Laboratory, ICMR Centre, V.S.S. Medical College, Burla, Sambalpur from May 1988 to June 1989. A total number of 1426 pregnant subjects were screened for sickling test in peripheral blood. The prevalence of sickle cell trait (AS) and sickle cell disease (SS) were 101.7 per thousand and 7.7 per thousand respectively. Episodes of crises during last trimester and labour was observed in SS mother exclusively (72.75%). Overt infection in the form of pyelonephritis was observed in 36.4% SS Mothers, Toxaemia was found to be high for SS (27.3%) & AS (17.24%) Mothers. Microscopic haematuria was observed in 27.3% of SS and 17.2% AS Mothers. Perinatal mortality was high in SS group 45%. No maternal death was encountered in this series.

Introduction :

"A disease more dreaded than Cancer" - was the comment on sickle cell disoder by Nalbadian etal (1972) because of its wide-spread and multitudinous manifestations which may affect any organ or system in the body.

The influence of sickle cell disorder on reproduction is ill-understood as it is only in the last four decades that women with sickle cell disease have survived to child bearing age in any number. The first report of a successful pregnancy was reported in 1931 by Yater and Mollari. Most of the workers consider pregnancy and sickle cell disease to be important mutual hazard.

The behaviour of this disease and its influence in pregnancy have been observed by many groups of Obstetricians in Jamaica, Africa, USA but its outcome is less known in India especially in this part of the country where the sickle cell gene is widespread in the general population.

Material and Methods:

During this fourteen month period, 1426 patients attending antenatal clinic and admitted for safe confinement were subjected to sickling test (Fig -1); positive result further needed Agargel electrophoresis of haemoglobin in alkaline pH to differentiate sickle cell trait (HbAS) and disease (HbSS). In the present study, a total of 145 pregnant mothers with sickle cell trait (HbAS) and 11 pregnant mother with sickle cell disease (HbSS) were identified among 1426 subjects. No other haemoglobinopathy was associated with them. The outcome of pregnancies was observed in these patients and compared with control group (HbAA).

Observation :

Table - 1 depicts the prevalence of sickle cell gene in the present study which was found to be 10.94% of all pregnancies of which the sickle cell disease was 0.77 percent and trait was 10.17%. Table - II Outlines the age, gravidity and parity of sickle cell trait, disease and control group. Associated complications during present pregnancy are outlined in Table - III.

Table I

Prevalence

Type of Patients	No. of Cases	Percent
Sickle cell disease (SS)	11	0.77
Sickle cell trait (AS)	145	10.17
Control (AA)	1270	89.06

Acute episodes of crises in the form of bone and joint pain in last trimester of pregnancy and labour of SS mother was the most striking complication. The other complications which were found to be high urinary tract infection in the form of Pyelonephritis (SS-36.4%, AS-6.9%), toxaemia (SS-27.3%, AS-17.24%) and

Table II

Age, Gravidity and Parity

	SS Group	AS Group	AA Group
Average age in years	24.5	23.8	23.8
Range	16-38	16-35	16-38
Average Gravidity	2.9	2.1	2.3
Range	1-6	1-6	1-7
Average Parity	1.3	1.6	2.1
Range	1-2	1-5	1-6
% of primigravida	45.4	41.3	40

Table - III

Associated Complications during Pregnancy

	SS		AS		AA	
	(n=11)	%	(n=145)	%	(n=1270)	%
1. Sickle Cell Crisis						
Vaso-occlusive						
Bone Pain	3	27.3	-	-		
Joint Pain	5	45.45	-	-		
2. Haemolytic	-	-	-			
Overt infection						
Respiratory	1	9.1	-	-	8	0.64
Urinary	4	36.4	10	6.9	25	1.98
3. Toxaemia	3	27.3	25	17.24	58	4.23
4. Cardiac failure	1	9.1	-	-	8	0.64
5. Thrombo embolism	1	9.1	-	-	6	0.49
6. Anaemia						
Moderate	9	81.8	70	48.3	309	24.36
Severe	2	18.2	-	-	179	13.60
7. Microscopic						
haematuria	3	27.3	25	17.2	63	5

Microphotograph of sickling of Red cells in pregnant mothers

microscopic haematuria among sickle cell mothers. Moderate to severe anaemia was marked in pregnancy of SS groups.

Labour was sponteneous in majority of the patients. Most deliveries were by vaginal route; for SS mother 81%, for AS mother 78%, for AA Control 76% respectively. Most of the babies of SS mothers were low birth weight, but there was no significant difference in birth wt. of AS mother and control group. Post-partum haemorrhage was noted in 18.18% in SS group. Blood transfusion was required more frequently in SS group. Perinatal mortality was high in SS group (45%) No maternal death occured among sickle cell mother in this series.

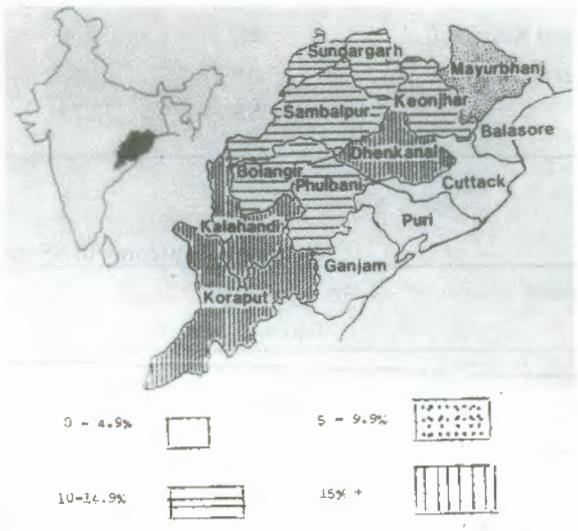

Fig : 2 Map of Orissa with its 13 Districts: sickle Cell trait prevalence

Discussion :

The prevalence of sickle cell disorder was observed to be high in Western Orissa i.e. 10-15% in general population as per study sickle cell research laboratory ICMR in this Institution (Kar et al 1978). Fig. II.

Assuming that the women in reproductive age group constitutes 25% of the population, the prevalence of sickle cell disorder should be 3.5%. It was also observed in the study of sickle cell research laboratory that for every 18 cases of trait (HbAS) one sickle cell disease (HbSS) is expected in this Zone. But the observed value for SS mothers was 0.77% instead of 0.19% i.e. four times higher.

Table - IV

Complications during Pregnancy reported in other study

Authors	Category of Patients	Crisis (%)	UTI (%)	Increase anaemia (%)	Toxaemia (%)	Microscopic haematuria (%)	Pneumonia %
Adams et al 1953, USA	S.C. trait	exact incidence not stated	-	-	24.2	-	-
	S.C. Anaemia	-	-	-	40	-	-
Anderson et al 1960 Jamaica	SS	88 Bone & Jt. Pain	-	-	22.2	11.1	-
Mc Curdy	AS	-	6.3	-	17.9	-	1.9
1964 USA	SS		10	20	5	-	25
Freeman & Ruth 1969 USA	SS	-	33	-	-	-	-
Hendricks et al, 1972, Nigeria	SS	68 Bone Pain	7	73	5	-	5
Tuck et al 1983	AS	-	16	-	-	16.5	Increase
United Kingdom	SS	38.4	28	-	12	13.6	
Present Study	AS	-	6.9	-	17.24	-	
	SS	72.75	36.4	56.51	27.3		9.1

Table - V

Pregnancy outcome in SS mothers reported in literature since 1956

Authors	No. of patients Patients	Total Pregnancies	Spontaneous abortions	Still birth	Neonatal death	Maternal death	Perinatal mortality	Fetal loss %
Abram & Schwartz 1959 Philadelphia, USA	6	30	8	1	0	0	4.5	30
Anderson et al, 1960 Jamaica	9	17	1	2	0	0	12.5	17.8
McCurdy, 1964 Washington D.C. USA	19	52	18	-	-	1	-	35
Hendricks et al 1972 Nigeria	38	4	11	4	8	7	18.2	18
Freeman & Ruth, 1969 Georgia, USA	18	41	11	4	1	0	15.1	37
Pritchard et al 1973 Texas, USA	34	50	16	5	4	0	25.0	42
Present study	11	31	13	5	4	0	29.03	57.6

This can be explained as the present study was undertaken in the referral hospital; the high prevalence of SS pregnant mother may be due to selection of as patient in hospital practice.

Adams et al (1953) reported incidence of sickle cell trait (7.9%), for disease 0.04%, in pregnancy. Anderson et al (1960) in Jamaica noted the incidence of sickle cell disease with Pregnancy to be 0.33%. In Mc Curdy's Series (1964) it was 6.4% for trait, 0.09% for disease. Hendricke et al (1972) had studied 38 patients of sickle cell disease in a span of 12 years.

Table IV Outlines the complications during pregnancy

of sickle cell mothers by various authors. In these different studies, besides acute episodes of crisis, Toxaemia, urinary tract infection and microscopic haematuria were observed frequently.

Adams et al (1953) questioned whether the incidence of toxaemia in sickle cell mothers was actually increased or the symptoms were produced by the disease itself. These worker were of the view that both the diseases were considered to be diseases of vascular system. Anoxia is due to vasospasm in toxaemia whereas anaemia and stasis due to sickle cell leads to anoxia and from this point the process is markedly similar.

Blank et al(1969) attributed the increased incidence of Pyelonephritis in sickle cell mother is related to scarring in renal Medulla which predisposes to infection. This scarring is due to repeated sickling of red cells in Vasarecta Urinary stasis of pregnancy aggravates infection. Increased incidence of haematuria noted in various studies can be attributed to renal medullay infarction and papillary necrosis.

Table VI depicts the outcome of pregnancy in SS mother reported by other workers in literature. It has been suggested that the presence of abnormal haemoglobin syndrome in a gravid state adversely affects foetal survival.

Foetal loss was high even when adequate antenatal Care was given.

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