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Sickle Cell Disease in Pregnancy: Maternal and Fetal Outcome in Port Harcourt, Nigeria

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Authors' contributions

This work was carried out in collaboration between all authors. Author IOG designed the study, wrote the protocol, and wrote the first draft of the manuscript. Author HAAU managed the literature searches, analyses of the study. All authors read and approved the final manuscript.

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ABSTRACT

Background: Medical experts for many years have daunted the occurrence of pregnancy in homozygote sickle cell patients. This is because of associated high risk for mother and fetus.
The aim of this study is to determine the prevalence and maternal and fetal outcome of pregnant mothers with sickle cell disease at the University of Port Harcourt Teaching Hospital, Nigeria.
Materials and Methods: This was a retrospective descriptive study of medical case files of all booked pregnant mothers who attended the antenatal clinic of the University of Port Harcourt Teaching Hospital, Nigeria from January 2007 to December 2011. The parameters extracted from the folders included: age, level of education, hemoglobin genotype, full blood count, malaria parasite, urine analysis and culture, complications of pregnancy, Apgar scores and birth weight.
Results: A total of 4,650 mothers were booked for antenatal care. Eight hundred and forty (18.1%) of them were HbAS, five (0.1%) were HbAC, nine (0.2%) were HbSS and 1(0.02%) HbSC. Age and gestation at booking were 18–42 years (mean 28.6± 2.1) and 9–34 weeks gestation (mean

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16.6±3.3), respectively. Malaria and vaso-occlusive crisis were the commonest complications encountered in pregnancy. Twenty percent of women had induction of labour and 60% were delivered by emergency caesarean section. Twenty percent had postpartum haemorrhage. Forty four percent of women delivered before 37 completed weeks. Birth weight below 2500 g occurred in 50% of singleton pregnancies. Two neonates developed transient complications related to maternal opiate exposure postnatally. There were 2(20%) maternal and fetal losses from toxaemia of pregnancy.

Conclusion: Pregnancy is uncommon among females with sickle cell disease in Port Harcourt, Nigeria. Sickle cell disease remains a severe complicating factor to pregnancy and associated with increased fetal and maternal losses.

Keywords: Sickle cell disease; pregnancy; maternal and fetal outcome.

1. INTRODUCTION

Sickle cell disease (SCD) comprises a group of inherited red blood cell conditions that result from the synthesis of variant or mutant haemoglobins [1]. SCD originates in tropical regions as a result of its advantage against malaria [1]. It is predominant among people from African, Asian, Arabian and Mediterranean countries; however, it is a worldwide health problem because of population migration [2]. SCD results in early childhood death if left untreated, and its effect on the burden of health care is being recognized as a global issue in terms of chronic disease.

The sickle mutation (β^s) of the beta-globin locus (HBB) is associated with five "classical" haplotypes [3]. These are named according to their putative geographical origins – Benin, Bantu (Central African), Cameroon, Senegal and Arab. The Benin haplotype is very common (~92%) among the Yoruba in Nigeria [3].

It is a disease of great clinical variability. It is only in the last half of the 20th century that women with SCD have survived to reproduce [4]. Early experience with SCD and pregnancy was a cause for condemnation, and the first report of a successful pregnancy was only in 1931 [4]. The first major review, in 1941, reported a 50% fetal loss [4]. Very high maternal mortality rates of 11.5% have been reported from West Africa and from black American groups [5]. These high mortality rates have led to suggestions by medical experts that pregnancy in SCD patients is contraindicated and have advised therapeutic abortion and sterilization [4-6]. The initiation of early aggressive prenatal care has dramatically improved perinatal outcome and reduced maternal mortality to less than 1% [7].

In view of that, there have been a number of observational reports on maternal mortality rates and fetal outcome in Africa and some parts of Nigeria [8,9]. There is no documented evidence on prevalence of sickle cell disease among pregnant mothers in our centre, Port Harcourt, Nigeria.

The present study was undertaken to assess the prevalence and pregnancy outcome of SCD mothers in Port Harcourt, Nigeria. The result obtained will be used to improve management of sickle cell diseases in pregnancy and reduce the burden of the disease in Port Harcourt, Nigeria.

2. MATERIALS AND METHODS

This was a retrospective cohort study of pregnant women who attended the antenatal clinic of the University of Port Harcourt Teaching Hospital (UPTH), Port Harcourt, Nigeria from Jan 2006 to December 2011. The UPTH is located in Port Harcourt metropolis, the heart of the Niger Delta region of Nigeria. The topography is that of flat plains with a network of rivers, tributaries and creeks which have a high potential for breeding of mosquitoes.

The information about each pregnancy was collected from the medical case notes of the patient which include haemoglobin genotype, full blood count, malaria parasite, urine analysis and culture, gestational age at delivery, mode of delivery, delivery outcome, birth weight, blood loss at delivery, coexisting medical conditions and complications during pregnancy and labour. Gestational age of the fetus was calculated from the first day of the last menstrual period and/or ultrasound dating. All the mothers at booking were routinely investigated for VDRL, HIV, Hepatitis B surface antigen, full blood count, blood group and genotype. Futhermore, malaria prophylaxis in the form of intermittent preventive therapy with fansidar is usually administered after 16 weeks and before 36 weeks. However,

for those with sickle cell disease, the following additional measures were taken. They were seen more frequently, two weekly till 28 weeks, and then weekly till delivery. Intermittent preventive therapy is usually given three times, at 20, 28 and 35 weeks gestation. In addition full blood count, blood film for malaria parasite and reticulocyte count were done at booking, 28, 36 weeks, and during labour.

Clinical diagnosis of a painful episode was made based on the subjective complaint of pain, overall clinical suspicion, and exclusion of other etiologies of pain based on associated symptoms.

Hemoglobin electrophoresis was done on cellulose acetate in alkaline buffer, using Helena's Electrophoresis System and staining by Ponceus stain. Scanning with Clinician 2 (Helena Laboratories Densitometer) was used for quantitative evaluation of different hemoglobin bands.

Hemoglobin electrophoresis which was carried out without history of blood transfusion during the previous four months was taken into consideration.

Malaria was defined as detection of malaria parasites in thick or thin peripheral blood films in the laboratory in any person (symptomatic or asymptomatic) [10]. Vaso occlusive crisis is painful crisis seen in patients with sickle cell disease [1]. Hyper-haemolytic crisis is defined as massive haemolysis with severe anaemia associated with sickle cell disease [1]. Preeclampsia was defined as a systolic pressure of 140 mm Hg or greater or a diastolic pressure of 90 mm Hg or greater after 20 weeks of gestation in the presence of proteinuria in mothers who were not chronically hypertensive [8]. While, eclampsia is an acute and life-threatening complication of pregnancy characterized by the appearance of tonic–clonic seizures (convulsions), usually in a woman who has developed pre-eclampsia [8]. Data were analyzed by the Statistical Package for the Social Sciences (SPSS for windows version 15.0, SPSS Inc.).Descriptive statistics in the form of the frequency and percentage were calculated.

3. RESULTS

A total of 4,650 mothers were booked for antenatal care. Eight hundred and forty (18.1%) of them were HbAS, five (0.1%) were HbAC, nine (0.2%) were HbSS and 1(0.02%) HbSC. Age and gestation at booking were 18–42 years (mean 28.6±2.1) and 9–34 weeks gestation (mean 16.6±3.3), respectively. Of the mothers with SCD, about 6 (60%) of were primiparous and 4(40%) were multiparous. Also, vaso-occlusive crisis in the form of bone pains was the most frequently encountered crisis in them preconception. Previous blood transfusion was also observed in 5 of the mothers. Malaria and vaso-occlusive crisis were the commonest complications encountered in mothers with sickle cell disease (Table 2). Twenty percent of women had induction of labour and 60 % were delivered by emergency caesarean section (Table 1). Twenty percent had a postpartum haemorrhage. Forty percent of them were delivered before 37 completed weeks (Table 3). Birth weight below 2500 g occurred in 50% of singleton pregnancies. Of these, two (40%) of them were delivered by mothers with preeclampsia. Two neonates developed transient complications related to maternal opiate exposure postnatally. There were 2(20%) maternal and fetal losses at 32 and 34 gestational ages (by emergency caesarian section) from toxaemia of pregnancy ($p=<0.05$).

4. DISCUSSION

We found only ten (0.2%) pregnant mothers with sickle cell disease. This is rather small compared to previous Nigerian studies [8,9]. This may mean that many of the affected children die before reaching child bearing age. Also, relative infertility among females with sickle cell disease may play a role as acknowledged in other studies [11,12].

Table 1. Socio-demographics of the pregnant mothers with Sickle cell disease (n-10)

Age in yrs (%)	Level of education (%)	G A at booking in weeks (%)	Parity (%)	Mode of delivery (%)
15-19= 3(30)	Primary= 0(0)	0-12= 3(30)	1= 4(40)	SVD= 4(40)
20-24= 4(40)	Secondary= 2(20)	13-28= 6(60)	2= 5(50)	C/S= 6(60)
25-39= 2(20)	Tertiary= 8(80)	29-36=1(10)	3= 1(10)	
30-35= 1(10)				

Key: yrs= years; GA= Gestational Age

Table 2. Complications among pregnant mothers with sickle cell disease (n=10)

Complications	Number	Percentage
Malaria	8	80
Vaso-occlusive crisis	5	50
Hyperhaemolytic crisis	3	30
Toxaemia of pregnancy	2	20
Urinary tract infection	2	20

Table 3. Fetal outcome of pregnant mothers with sickle cell disease (n=10)

Fetal outcome	Number	Percentage
Still birth	2	20
Birth asphyxia	2	20
Prematurity	4	40
Low birth weight	5	50

It has been documented that pregnant women with sickle cell disease have more frequent crises due to the extra stress of pregnancy [6]. This is because pregnancy is an intense burden on a woman's body, and this incredible strain can easily exacerbate the sickling of red blood cells. When these cells cluster together, they can build up in various organs throughout the body, leading to intense pain. In this study half of the pregnant mothers had vaso-occlussive crisis which was precipitated by infections due to malaria. This finding had been documented by previous Nigerian studies [13,14].

Malaria is widely considered a major cause of illness and death in patients with SCA in sub-Saharan Africa [10], but the evidence to support this has been conflicting. Some researchers have reported an association between malaria and admission to hospital, anaemic crises [15], and mortality [16]; however, these studies were done in areas of high malaria transmission where the finding of the anaemia cannot be taken as proof that malaria was cause of the clinical presentation.

Pregnancy in SCD patients is linked with increased risk to both mother and fetus. Several studies have reported very high maternal mortality rates in Africa and from black American groups [17-18]. These high mortality rates have led to propositions by clinicians that pregnancy in SCD patients is contraindicated and have advised therapeutic abortion and sterilization [4]. In our study we found 20% rates of fetal and maternal losses late in pregnancy which is similar to that reported in Jamaica [19]. The limited laboratory facilities in our hospital have hindered attempts to investigate the causes of these fetal deaths in more detail. These deaths might have been prevented by inducing labour earlier.

Low birth weight is one of the most consistent finding in infants born to mothers with SS disease [20]. Half of the infants in our study were low birth weight. Several factors such as prematurity, preeclampsia, acute anaemic episodes, and the number of SS-related complications during pregnancy may be responsible for the low birth weight in mothers [20,21].

Preeclampsia was responsible for the maternal deaths in our study. This is at variance with a Saudi Arabian study where none (of pregnant mothers with sickle cell disease) had preeclampsia [18]. The mechanism for preeclampsia remains unclear; multiple factors such as placental ischaemia and endothelial injury have been implicated. It can be treated with bed rest at home or in the hospital, if needed. Delivery of the fetus may be required, if the preeclampsia deteriorates, at gestational age greater than 32 weeks.

5. CONCLUSION

The current study suggests that there is high maternal and fetal mortality in pregnant mothers with sickle cell disease. A multidisciplinary team approach to deal with antenatal complications in SCD women will improve pregnancy outcome in these patients.

CONSENT

It is not applicable.

ETHICAL APPROVAL

Ethical approval was obtained from the Ethical Committee of the University of Port Harcourt Teaching Hospital, Nigeria.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

REFERENCES

1. Serjeant GR. The emerging understanding of sickle cell disease. British Journal of Haematology. 2001;112:3-18.
2. Stuart MJ, Nagel RL. Sickle-cell disease. Lancet. 2004;364:1343-60.
3. Flint J, Harding RM, Boyce AJ, Clegg JB. The population genetics of the haemoglobinopathies. Baillieres Clin Haematol. 1998;11:1-51.
4. Oteng-Ntim E, Cottee C, Bewley S, Anionwu EN. Sickle cell disease in pregnancy. Current Obstetrics and Gynaecology. 2006;16:353–60.
5. Sun PM, Wilburn W, Raynor BD, Jamieson D. Sickle cell disease in pregnancy: twenty years of experience at Grady Memorial Hospital, Atlanta, Georgia. American Journal of Obstetrics and Gynecology 2001;184:1127-30.
6. Villers MS, Jamison MG, De Castro LM, James AH. Morbidity associated with sickle cell disease in pregnancy. Am J Obstet Gynecol. 2008;199(2):125.
7. Sonawane AS, Zodpey SP. Pregnancy outcome in sickle cell disease/trait. J. Obstet Gynaecol India. 2005;55:415-418.
8. Ocheni S, Onah HE, Ibegbulam OG, Eze MI. Pregnancy outcomes in patients with sickle cell disease in Enugu, Nigeria. Niger J Med. 2007;16(3):252-5.
9. Idrisa A, Omigbodun AO, Adeleye JA. Pregnancy in hemoglobin sickle cell patients at the University College Hospital, Ibadan. Int J Gynaecol Obstet. 1992;38:83-6.
10. Idrisa A, Omigbodum AO, Adeleye JA. Pregnancy in hemoglobin sickle cell patients at the university college hospital, Ibadan. Int J Gynecol Obstet. 1992;38:83-6.
11. Serjeant GR, Hambleton I, Thame M. Fecundity and pregnancy outcome in a cohort with sickle cell-haemoglobin C disease followed from birth. BJOG. 2005;112:1308-1314.
12. Afolabi BB, Iwuala NC, Iwuala IC, Ogedengbe OK. Morbidity and mortality in sickle cell pregnancies in Lagos, Nigeria: a case control study. J Obstet Gynaecol. 2009;29:104-106.
13. Dare FO, Makinde OO, Faasuba OB. The obstetric performance of sickle cell disease patients and homozygous hemoglobin C disease patients in Ile-Ife, Nigeria. Int J Gynecol Obstet. 1992;37:163-8.
14. Odum CU, Anorlu RI, Dim SI, et al. Pregnancy outcome in Hb SS cell disease in Lagos, Nigeria. West Afr J Med. 2002;21:19-23.
15. Ashley-Koch A, Young Q, Olney RS. Sickle haemoglobin (HbS) allele and sickle cell disease: a HuGE review. American Journal of Epidemiology. 2000;151(9):839-45.
16. Ikefuna AN, Emodi IJ. Hospital admission of patients with sickle cell anaemia pattern and outcome in Enugu area of Nigeria. Nigerian Journal of Clinical Practice. 2007;10(1):24-29.
17. Hendrickes JP, Harrison KA. Pregnancy in homozygous sickle cell anaemia. Br J Obstet Gynecol. 1972;79:396-409.
18. Khalifa AM. Pregnancy in sickle cell disease in the Al Hassa region of Saudi Arabia .Annals of Saudi Medicine. 2000;20(5&6):471-473.
19. Poddar D, Maude GH, Plant MJ, et al. Pregnancy in Jamaican women with homozygous sickle cell disease, fetal and maternal outcome. Br J Obstet Gynaecol. 1986;93:727-32.
20. Serjeant GR, Look Loy L, Crowther M, Hambleton IR, Thame M. Outcome of pregnancy in homozygous sickle cell disease. Obstet Gynecol. 2004;103:1278-1285.
21. Smith JA, Espeland M, Bellevue R, Bonds D, Brown AK, Koshy M. Pregnancy in sickle cell disease: experience of the cooperative study of Sickle cell disease. Obstet Gynecol. 1996;87:199-204.

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