

Original Research Article

Sickle Cell Disease in pregnancy: Maternal and fetal Outcome in Port Harcourt, Nigeria

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 pregnancy outcome of mothers with sickle cell disease at the University of Port Harcourt Teaching Hospital, Nigeria.

Materials and methods: This was a retrospective 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 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 a postpartum haemorrhage. Forty four percent of women delivered before 37 completed weeks. Birth weight below 2500 g occurred 50% of singleton pregnancies. Two neonates developed transient complications related to maternal opiate exposure postnatally. There were 2(20%) maternal and fetal losses from toxemia 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. Therefore, a strategy of exchange blood transfusion of all women with homozygous sickle cell disease (HbSS) from 28 weeks gestation is recommended to reduce the risk of maternal complications in the third trimester and puerperium.

Key Words: Sickle Cell Disease; Pregnancy; maternal and fetal outcome.

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.

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[3]. Early experience with SCD and

pregnancy was a cause for condemnation, and the first report of a successful pregnancy was only in 1931[3]. 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 [4]. 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[3-5]. The initiation of early aggressive prenatal care has dramatically improved perinatal outcome and reduced maternal mortality to less than 1%[6].

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. 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 and reduce the burden of the disease in our locality.

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, Port Harcourt, Nigeria from Jan 2006 to December 2011.

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 last menstrual period and/or ultrasound dating. 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.

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. 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 (Table1). Twenty percent had a postpartum haemorrhage. Forty percent of them were delivered before 37 completed weeks (Table3). Birth weight below 2500 g occurred 50% of singleton pregnancies. Two neonates developed transient complications related to maternal opiate exposure postnatally. There were 2(20%) maternal and fetal losses from toxemia of pregnancy.

DISCUSSION

We found only ten (0.2%) pregnant mothers with sickle cell disease. This is rather small compared to previous Nigerian studies[7,8]. This may mean that many of the infected

112 children die before reaching child bearing age. Also, relative infertility among females
113 with sickle cell disease may play a role as acknowledged in other studies[9,10].

114 It has been documented that women who are pregnant with sickle cell disease have
115 more frequent crises due to the extra stress of pregnancy[5]. This is because pregnancy
116 is an intense burden on a woman's body, and this incredible strain can easily
117 exacerbate the sickling of red blood cells. When these cells cluster together, they can
118 build up in various organs throughout the body, leading to intense pain. In this study half
119 of the pregnant mothers had vaso-occlusive crisis which was precipitated by infections
120 due to malaria. This finding had been documented by previous Nigerian studies[11,12].

121 Malaria is widely considered a major cause of illness and death in patients with SCA in
122 sub-Saharan Africa[13], but the evidence to support this has been conflicting. Some
123 researchers have reported an association between malaria and admission to hospital,
124 anaemic crises[14], and mortality[15]; however, these studies were done in areas of
125 high malaria transmission where the finding of parasitaemia cannot be taken as proof
126 that malaria was the cause of the clinical presentation.

127 Pregnancy in SCD patients is linked with increased risk to both mother and fetus.
128 Several studies have reported very high maternal mortality rates in Africa and from
129 black American groups[16-17]. These high mortality rates have led to propositions by
130 clinicians that pregnancy in SCD patients is contraindicated and have advised
131 therapeutic abortion and sterilization[3]. In our study we found 20% rates of fetal and
132 maternal losses late in pregnancy which is similar to that reported in Jamaica[18].The
133 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[19]. Half of the infants in our study were low birth. Several factors such as prematurity, preeclampsia, acute anemic episodes, maternal fetal haemoglobin (HbF) levels, and the number of SS-related complications during pregnancy may be responsible for the low birth weight in mothers[19,20].

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[17].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.

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. Therefore, a strategy of exchange blood transfusion of all women with homozygous sickle cell disease (HbSS) from 28 weeks gestation is recommended to reduce the risk of maternal complications in the third trimester and puerperium.

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206 **Table 1: Socio-demographics of the pregnant mothers with Sickle cell disease (n-**
207 **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= 4(40)	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)				

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209 **Key: yrs= years; GA= Gestational age**

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213 **Table 2: Complications among Pregnant Mothers with Sickle Cell Disease (n=10)**

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

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224 **Table 3: Fetal Outcome of Pregnant Mothers with Sickle Cell Disease (n=10)**

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

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