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Full Length Research paper

Feto-maternal outcome in patients with sickle cell anaemia at the University of Port Harcourt Teaching Hospital, Port Harcourt, Nigeria: A ten year retrospective review

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Sickle cell anaemia is associated with adverse feto-maternal outcome especially in less resource countries with poor obstetrics care. The objective of this study was to evaluate the feto-maternal outcome in pregnant women with sickle cell anaemia at the University of Port Harcourt Teaching Hospital, Port Harcourt. A retrospective review of 17 pregnancies in 13 patients with sickle cell anaemia over a 10-year period was conducted at the University of Port Harcourt Teaching Hospital, Port Harcourt between January 2003 and December 2012. The data retrieved from the case notes of the patients were analysed using SPSS version 17.0. The mean age of the women with sickle cell anaemia reviewed was 29.3 years. Most (76.5%) of them were booked and delivered at term (70.6%). All the women were anaemic out of which 9 had blood transfusion. Other complications ranged from vaso occlusive crises (100%), through urinary tract infection (64.8%), to lobar pneumonia (17.7%) and hypertensive disorders (17.7%). A maternal mortality rate of 9.7 per 100,000 deliveries was found. The mean birth weight was 2752.9g. The major fetal complications were neonatal jaundice (47.1%) and neonatal sepsis (23.5%).The stillbirth rate was 0.1 per 1000 live births. Sickle cell anaemic patients are at increased risk of adverse feto maternal outcomes in pregnancy especially in less resource centres and therefore should be managed in centres with adequate personnel and facilities.

Key words: Sickle cell anaemia, Vaso-occlusive crisis, Preterm delivery, feto-maternal outcome, Complications**.**

INTRODUCTION

Sickle cell disease (SCD) remains the commonest genetic disease worldwide and includes disorders affecting the structure and function of haemoglobin (Ohl and Christensen, 2004). It is common among the black race with the highest incidence in the world reported from Uganda (Lawson, 1967), in East Africa. In Nigeria as reported by Alolabi et al (2009), about 25% of the entire population have the sickle cell trait and approximately 100,000 children are born annually with a serious sickle cell disorder. Haemoglobin SS (HbSS) is the most common pathological variant worldwide with such patients suffering sickle cell anaemia (SCA) (Weatherall et al 2006). As illustrated by Ocheri et al, (2007) and Ngoh et al, (2004), it occurs when there is a substitution of the amino acid valine for glutamic acid at position 6 of the i-chain leading to a gross reduction in the lifespan of red blood cells from the expected 120 days in the unaffected population to 10-20 days, with the sequel of chronic anaemia in the affected patients.

There is an increase in the number of women who survive into adulthood and get pregnant attributable to improvement in medical care. This has led to an increasing prevalence of sickle cell anaemia in pregnancy in recent times, making it a disorder of public health concern in today's obstetrics (Omo-Aghoja, Okonofua, 2007; Sowane, Zodpey 2005 andSerjeant et al ,2004).

Pregnant women with this genetic disorder manifest some complications with associated fetal morbidities. The

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early reports (Odum et al, 2002; and Dare et al, 1992) on the outcome of pregnancy in women with sickle cell anaemia depicted an almost universal adverse outcome for mother and child. The advances however in medical care including the introduction of pre-conception care has shown greatly improved feto maternal outcomes among these women (Omole-Ohonsi et al, 2012; Serjeant et al, 2004). On the other hand, (Omo-Aghoja and Okonofua, 2004; Sowane and Zodpay, 2005; Dare et al, 1992; Al Kahtani et al, 2012) have documented varying incidences of maternal and neonatal complications in developed and developing countries. Countries with improved healthcare delivery systems have documented a reduction in maternal mortality associated with sickle cell anaemia with consistently reduced rates as years go by while Sub-Saharan countries such as Ghana and Nigeria (Dare et al, 1992; Ratimy et al, 2000) have a high prevalence of SCA and maternal mortality rates exceeding 9%.

Documented complications as cited by Afolabi et al, (2009); Odum et al, (2002) and Omole-Ohonsi et al (2012); range from pre-partum, intra-partum and postpartum painful crises, pulmonary complications, anaemia, hypertensive disorders, premature delivery with its attendant risks, increased operative deliveries and intrauterine growth restriction.

There exist scanty literature on sickle cell anaemia in pregnancy from Sub-Saharan Africa; with available studies (Dare et al, 1992; Afolabi et al, 2009 and Ratinmy et al, 2000) suggesting maternal mortality varying from 1.8% to 9%. Given the paucity of information currently available on feto maternal outcomes, reviewing the maternal and neonatal complications in this study will give an insight to improved management. Hence this study evaluated the feto maternal outcomes among women with sickle cell anaemia in Port Harcourt, Nigeria.

MATERIALS AND METHODS

A 10-year retrospective study of all cases of pregnant women with sickle cell anaemia between January 2003 and December 2012 was carried out. Records of the antenatal clinic, antenatal ward, labour ward, theatre and neonatal care unit of pregnant patients with sickle cell anaemia were retrieved. The research ethics committee of University of Port Harcourt Teaching Hospital gave due ethical approval.

A total of 13 women with HbSS who had 17 pregnancies were studied. They were managed according to the department's protocol for pregnant women with SCA. This included frequent antenatal visits (fortnightly antenatal clinic visits till the third trimester, then weekly visits), investigating for and treatment of infections including malaria at each visit, prevention and prompt treatment of specific complications such as crisis, and blood transfusion with haemoglobin AA blood, when clinically indicated and/or when the haemoglobin concentration drops to 6 g/dl or below. The protocol also included regular antenatal fetal monitoring, intensive intrapartum and postpartum care of the women and their babies. Due to its location and the peculiarity of this centre, unbooked patients and referred patients were included in this study.

The relevant data extracted and placed on a spread sheet included socio-demographic variables, booking status, gestational age at booking, packed cell volume estimation (at booking, during pregnancy, delivery and puerperium), number of antenatal visits and complications during the antenatal period, blood transfusion during pregnancy (if any), gestational age at delivery, mode of delivery and postpartum complications. Data on fetal outcome was also extracted.

The data were analysed using SPSS version 17.0 (IBM, Armonk, NY, USA). The results are presented as percentages for frequencies and as mean and standard deviations for continuous variables.

RESULTS

Study population

There were 30,934 deliveries including twin and triplet births at the Department of Obstetrics and Gynaecology of the University of Port Harcourt Teaching Hospital from 2003 to 2012 of which 17 (0.05%) were from 13 women with SCA. Four of the women had two pregnancies during the study period, thus a total of 17 pregnancies were studied.

Table 1 shows the age distribution of the women studied ranged between 25 to 38 years with a mean age of 29.3 years. Eight (47.1%) of the pregnancies were in nulliparous women while 3 (17.7%) were in multiparous women. All the women were married and had some form of formal education. 12(70.6%) of the women had attained tertiary level of education. Most (76.5%) of the women were booked. All deliveries occurred above 30 weeks gestational age with 12 (70.6%) having delivered at term. The rest were preterm deliveries between 31 and 36 weeks of gestation.

Table 2 shows the major complications that were present among the women.

They were all anaemic. Majority of them had packed cell volumes estimation between 21 and 23% at the time of delivery. Nine patients required blood transfusion in the antepartum period with 3 receiving multiple blood transfusions. All 17 pregnancies in the study group were complicated by vaso-occlusive crises with one of them having haemolytic crises. Eleven patients (64.8%) had urinary tract infection (UTI) while 3 (17.7%) had lobar pneumonia. There were 2 patients who had both infections. Osteomyelitis occurred in one patient. Hypertensive disorders complicated 3 of the pregnancies.

There were 3 deaths among the women with SCA during the study period giving a mortality rate of 9.7 per 100,000

Table 1. Socio-demographic characteristics of the study group.

Characteristics	Frequency	Percentage %
Age		
≤20	0	0
21 – 25	2	11.8
26 – 30	9	52.9
31 – 35	5	29.4
36 – 39	1	5.9
≥40	0	0
Parity		
0	8	47.1
1	6	35.3
2	2	11.8
3	1	5.9
4	0	0
≥5	0	0
Marital Status		
Married	17	100
Single	0	0
Educational status		
No formal education	0	0
Primary	0	0
Secondary	5	29.4
Tertiary	12	70.6
Occupation		
Student	2	11.8
Self-employed	4	23.5
House wife	6	35.3
Private sector	3	17.6
Public sector	2	11.8
Booking status		
Booked	13	76.5
Unbooked	3	17.6
Referred	1	5.9
GA at delivery		
28-30wks	0	0
31-36wks	5	29.4
37-42wks	12	70.6

deliveries.

Table 3 shows there were a total of 14 live births (82.4%) and 3 stillbirths in the study population with a stillbirth rate of 0.1 per 1000 live births (Mode of delivery was spontaneous vaginal in 2 and repeat caesarean section in 1). Two of these stillbirths were preterm. The birth weights of babies from the study group ranged between 1700g to 3600g with a mean birth weight of 2752.9g. Preterm delivery (gestational age at delivery < 37weeks) occurred in 5 women with the babies having low birth weight (LBW) defined as absolute birthweight< 2500g. (Of the 5 preterm deliveries, 3 were by the vaginal route while 2 were by emergency caesarean section. Among the 3 preterm babies delivered through the vaginal route, one was a stillbirth, one had neonatal jaundice and the 3rd had no complication while among the 2 preterm babies

Table 2. Maternal complications.

Complications	Frequency	Percentage %
Vaso-occlusive crises	17	100
Haemolytic crises	1	5.9
Anaemia	17	100
Urinary tract infection	11	64.8
Lobar pneumonia	3	17.7
Osteomyelitis	1	5.9
PIH	2	11.8
Pre-eclampsia	1	5.9
Haemolytic crises	1	5.9

Table 3. Fetal / Neonatal complications.

Complication	Frequency	Percentage %
Neonatal Jaundice	8	47.1
Neonatal Sepsis	4	23.5
Low birth weight (preterm)	5	29.4
Moderate birth asphyxia	1	5.9
Stillbirth	3	17.6

delivered by caesarean section, one was stillborn and the other had no complication.). One baby (5.9%) had moderate birth asphyxia, 8 (47.1%) had neonatal jaundice (NNJ) while 4 (23.5%) had neonatal sepsis (NNS). There was no early neonatal death (death within the 1st seven days after delivery).

DISCUSSION

Afolabi et al (2009); Serjeant et al (2004) and Yu et al, (2009) documented increased risk of maternal and fetal complications during pregnancy in sickle cell anaemic patients compared with healthy women. Although maternal and perinatal mortality rates have been on the decline over the years due to improvements in healthcare through multidisciplinary approaches, the trend is not the same worldwide. It is more evident in developed countries with improved healthcare delivery systems including pre-conception care.

This study was conducted in relatively young women with a mean age of 29.7 years, most of the women in the age range 25-31 years. This mean age was comparable to the mean age of 27.6 years in a similar study in Bahrain but with a better outcome than in this study. All the women in this study were married with adequate marital and family support. Majority had tertiary level of education with some form of employment (self, private or public). Thus, these women were significantly empowered to care for themselves and their pregnancies. This is reflected in about 76.5% of the pregnancies being booked in this tertiary health facility.

This study uncovered an increased risk of complications among women in Port Harcourt with SCA when compared with similar studies in Saudi Arabia and Bahrain (Al Kahtari et al, 2012 and Rajab et al, 2006). This difference might reflect improvements in the healthcare delivery as well as in the management of SCA in these countries when compared to Port Harcourt, Nigeria. It has also been attributed to the poor utilization of the scanty medical and antenatal care facilities as well as poor or non-existence of pre-conception care facilities in most communities in sub-Saharan Africa. Complications such as anaemia, haemolytic crisis and urinary tract infection were similar to those in the study in Ghana by Wilson et al (2012).

Anaemia in this study occurred in all the women. This was significantly higher than in some other studies by Al Khatari et al (2012) and Wierenga et al (2001), conducted in Saudi Arabia and Jamaica respectively. Better nutrition, pre-conception care as well as reduced infections may account for this observation. In 53% of the women, anaemia requiring antepartum blood transfusion was found. This was slightly higher than the rates of 40% and 45% by Afolabi et al (2009) and Odum et al (2002) within the same country. Much lower rates of antepartum

blood transfusion were documented in Saudi Arabia and Bahrain(Al Kahtari et al, 2012 and Rajab et al 2006). Prophylactic blood transfusion is recommended and practised as reported in certain studies (Yu et al, 2009 and Rajab et al, 2006) to reduce SCD-related complications of childbirth. This practice may however be less tolerated in developing countries where blood availability and transfusion has economic, cultural and religious bias. Randomized controlled trials however reveal no significant difference in the perinatal outcome between women with SCA who underwent prophylactic blood transfusion and those who did not as reported by Koshy et al (1988).

Vaso-occlusive crisis was the most common reason for hospital admission occurring in 100% of the women ranging from 1-3 episodes in the antepartum period. One woman also had an episode of haemolytic crises. Much lower rates were documented in studies by Odum et al, (2002) (41.4%); Al Kahtari et al (2012) (43.1%); and Rajab et al, (2006) (42.2%).Interestingly, the rate by Odum et al, (2002); in South-west Nigeria, is comparable to those outside the country. While the exact reason for this observation may not be known, possible explanations may be due to the higher incidences of infections as most of the crises were precipitated by infections. Less certain in this case is the well-known fact that Haemoglobin F (Hb F) whose level is raised in a form of SCD prevalent in the Middle East (Asian Beta-globin haplotype) plays a prophylactic role in reducing the incidence and severity of crisis as reported in Eastern province of Saudi Arabia (Al Kahtari et al, 2012) when compared to the Benin haplotype (African origin). None of these factors completely explains this observation. Thus, this observation may result from a combination of factors.

The rates of infections in this study were higher than in other previously compared studies. In a singular study by Yu et al (2009) a combined incidence of UTI, lobar pneumonia and osteomyelitis was at 19% in comparison to the high incidence in this study (82.5%).

Pre-eclampsia was lower than previously compared documented studies.This is at variance with the well-known higher incidence of pre-eclampsia among the Negroid race as reported by OmoleOhonsi et al (2012). Non-proteinuric hypertension was found in 11.8% of the patients in the study population.

SCA in pregnancy has been associated with Low birth weight (LBW) and has been observed in 28-42% of cases by Wierenga et al (2001). Factors responsible for this are varied and inconsistent but include lower gestational age at delivery, lower maternal age and weight, history of pre-eclampsia, anaemia and intrauterine growth restriction. In this study, 29.4% of the women delivered babies with LBW (< 2500g) and all were as a result of preterm labour. Other contributing factors to LBW include common interventions in the management of SCA in the form of early abdominal delivery or preterm induction of labour. The percentage of preterm delivery in this study is comparable to the 24% in a previous study by Yu et al (2009). The increased risk of LBW is associated with an increased risk of neonatal jaundice (NNJ) and birth asphyxia as was discovered in this study Infants born to 23.5% of the women had NNJ while 5.9% had moderate birth asphyxia.

As documented in the 1980s by Ngo et al, (1995) the perinatal mortality rate in the United Kingdom was high ranging between 40 and 60 per 1000 deliveries (4-6 times that of the normal population) with a maternal mortality rate of 2%. In Benin Republic, maternal and perinatal mortality rates of 27% and 40% respectively were reported Ratimy et al (2000). More recently however, a maternal mortality rate of 0.5% and a perinatal mortality rate of 77.7 per 1000 live births were observed in Saudi Arabia by Al Kahtami et al (2012).In this study, there were 2 maternal deaths and 3 cases of stillbirths giving maternal mortality rate and perinatal mortality rates of 9.7 per 100,000 deliveries and 0.1 per 1000 live births respectively. This illustrates the pattern as expected in Sub Saharan Africa and developing countries where the high maternal mortality is comparable.

Excessive maternal deaths can be prevented by improved health seeking behaviour, improved prenatal care and standard care of SCA in pregnancy. This has been reported in an African setting (Ratimy et al, 2000) presenting the possibility. In this study, the maternal deaths occurred in referred and unbooked patients who presented late for medical care and as such did not have adequate antenatal care.

CONCLUSION

Pregnant women with SCA experience an increased incidence of medical and pregnancy related complications. Nonetheless, medical advances in obstetrics and neonatology have improved feto-maternal outcome. This improved outcome is still not reflected in low resource settings due to multiple factors ranging from poor health seeking behaviour, poorly developed healthcare delivery systems in addition to socioeconomic, cultural as well as religious limitations.

It is recommended that public enlightenment, specialised multidisciplinary care and close monitoring in tertiary institutions be advocated to reduce these morbidities and mortalities associated with sickle cell anaemia in pregnancy.

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