PREGNANCY OUTCOME AMONG PATIENTS WITH SICKLE CELL DISEASE IN JOS, NORTH CENTRAL NIGERIA

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ABSTRACT

Background: *With advances in management, education, awareness and improved nutrition, men and women with sickle cell disease are enjoying an improved quality of life well into adulthood, when they elect to plan a family. As a result, sickle cell disease is a common haemoglobinopathy encountered during pregnancy in Nigeria. Reports from other parts of the country have documented increased maternal and perinatal morbidity and mortality, but none has been reported from Jos.*

Materials and Methods: *A retrospective review of pregnancy outcome in patients with sicle cell disease managed at the Jos University Teaching Hospital, Jos over a 5year period was carried out. Data extracted from patients' case files were analysed using simple statistical methods with Epi info 2002 statistical software.*

Results*: Case files of patients with sickle cell disease in pregnancy during the period under review were retrieved. The mean maternal age was 25.1±3.9 years. The mean gestational age at booking or first presentation was 19.3±7.7 weeks. The antenatal complications included anaemia (62.9%), vaso-oclusive (bone pain) crisis (58.1%), intra uterine growth restriction (45.7%), pregnancy induced hypertension and malaria 25.7% each. The maternal and perinatal mortality rates were 53 and 384 per 1000 live births respectively.*

Conclusion: *Pregnancy in sickle cell disease patients is associated with high maternal and perinatal morbidity and mortality as reported in other parts of the country. The importance of early presentation for antenatal care and a call for preconception care is made.*

Keywords: *Pregnancy, outcome, sickle cell, disease, Jos*

INTRODUCTION:

The term sickle cell disease refers to a lifelong disorder arising from the inheritance of two abnormal haemoglobin genes, each from a parent with at least one of the two abnormal genes being haemoglobin S. Examples of sickle cell disease are HbSS, HbSC and HbSB-thalasaemia.

In the deoxygenated state the HbS molecule forms polymers which lead to sickling of the red blood cells. The heterozygous form of sickle cell haemoglobinopathy (HbAS) is essentially a benign condition that is believed to have no particular antenatal sequelae. It confers partial protection from falciparum malaria.[1-3] The prevalence of sickle cell diseases is highest in Africans and people of Mediterranean or East Indian ancestry.[2,4,5]
With advances in management, education, awareness and improved nutrition, men and women with sickle cell disease are enjoying an improved quality of life well into adulthood, when they may elect to plan a family, as a result, sickle cell disease is a common haemoglobinopathy encountered during pregnancy in Nigeria.

There are, however, increased risks involved in pregnancy for the mother with sickle cell disease and her foetus. Maternal risks include exacerbated sickle cell disease phenomena such as anaemia, vaso-oclusion, and ischaemic injury as a result of the physiologic stress of pregnancy.[3] They have greater susceptibility to chest and urinary tract infections, hypertensive disorders of pregnancy and thrombo embolic complications.[2,5-7] Risks to the foetus include increased frequency of miscarriage,

intra uterine growth restriction and preterm labour and premature delivery. Perinatal morbidity and mortality are higher than in the general population.[2-6]Despite risks associated with sickle cell disease in pregnancy, appropriate management by health care providers familiar with sickle cell diseases and high risk obstetric care can result in successful pregnancy outcomes in patients with sickle cell diseases[3,5,6,8.] The pregnant woman suffering from sickle cell disease must be regarded as high risk.

Studies in Ibadan, Lagos, Enugu and other parts of southern Nigeria have all documented increased maternal and perinatal morbidity and mortality,[10,11,14] but there has been paucity of reports of pregnancy outcome in sickle cell disease from northern Nigeria. The objective of this study was to review the pregnancy outcomes in patients with sickle cell disease as seen at the Jos University Teaching Hospital, North Central Nigeria.

MATERIALS AND METHODS

This was a retrospective study of women with sickle cell disease who presented with pregnancy at the Jos University Teaching Hospital (JUTH) between 1st January 2003 and 31st December 2007. A thorough search of the antenatal, labour and delivery records of the obstetric unit as well as the records of the medical records department of the hospital was done to identify these patients. Their case files were retrieved from the Medical Records Department and studied.

Data extracted from the patients case files included: Age, parity, gestational age at booking, complications of the disease and pregnancy during the antenatal period, labour and puerperium. Information about mode of delivery and foetal outcome were also extracted from the case files. The data obtained was then analysed using simple statistical methods with Epi info 2002 statistical soft ware.

RESULTS

During the period under review, there were 13611 deliveries,and forty four women had sickle cell disease in pregnancy. Only the case files of 38patients with sickle cell disease in pregnancy could be retrieved. Thirty seven pregnancies were complicated by HbSS while one pregnancy was complicated by HbSC.

The mean maternal age was 25.05 ± 3.87 years. About 58% of the patients had secondary school education while 21% each had primary and tertiary education respectively.Over half (57.9%) of the patients were nullipara while none of the women had parity greater than three.

The mean gestational age at booking or first presentation was 19.3 +/- 7.7 weeks with only 25.7%(9) of the patients booking or presenting before 13weeks of gestation. Three women were referred in labour, having had there antenatal care elsewhere. The mean packed cell volume at booking was 26.3± 5.2%. About 68.9% of the patients required antenatal admission during the pregnancy due to maternal or foetal complications, with about 41.7% of these, requiring more than one antenatal admission during the pregnancy.

About sixty three (62.9%) percent of the patients had anaemia, with 31.4% presenting with severe anaemia (PCV ≤ 18%) or anaemic heart failure during the antenatal period. Bone pain crisis (51.4%), pregnancy induced hypertension (25.7%) and malaria (25.7%) were other common antepartum maternal complications observed. One patient with twin pregnancy had eclampsia at 32 weeks of gestation.

Intrauterine growth restriction was observed in 45.7% of the pregnancies while intrauterine foetal death occurred 20.5% of the patients, (Table 1). About forty two percent of the patients required antenatal blood transfusion; the mean number of units transfused was 3.3 pints (range 1-5).

The mean gestational age at delivery was 36.9 ± 2.29 weeks (range 32 – 41 weeks), with about 42.9% of the patients delivering before 37 completed weeks of gestation. The mean packed cell volume in labour was 24.07 ± 5.74 %, (range 15-34%). About seventy four percent (73.7%) of the pregnancies were delivered vaginally while 26.3% had caesarean section. The instrumental delivery rate was 26.3%, six women had vacuum and four had forceps delivery.The most common intrapartum complication observed in this series was anaemia (58.1%), other complications noted were: vaso occlusive crisis, foetal distress and failure to progress in labour due to cephalo pelvic disproportion (Table 2).

The mean birth weight was 2.39 ± 0.5kg, (range 1.3-3.2kg) with 69.2% of the babies weighing less than 2.5kg. The mean Apgar score at 5 minutes was 7.02 ± 2.24, (n=30). There were 15 perinatal deaths (38.4%). This was made of 8 (20.5%) stillbirth and 7

(17.9%) early neonatal deaths.

The commonest puerperal complication was anaemia (57.9%) with 36.8% requiring blood transfusion in the postpartum period. Other puerperal complications included bone pain crisis (10.5%), post caesarean section wound infection (5.3%). One patient had puerperal psychosis following a macerated stillbirth.About 28.9% of the patients had no puerperal complications, (Table 3).

There were two maternal deaths (5.3%), both occurring within seven days of delivery. The two maternal deaths had anaemia as a complication of their pregnancies, with one referred from a primary health care clinic in labour with anaemic heart failure.

Table 1: *Antenatal complications in women with sickle cell disease in pregnancy

Complication	N=35	%
Anaemia (pcv<30%)	22	62.9
Severe anaemia (pcv=18%) / anaemic heart failure	11	31.4
Vasooclusive crisis	18	51.4
Intrauterine growth restriction	16	45.7
Pregnancy induced hypertension/ Eclampsia	11	28.9
Malaria	9	25.7
Intrauterine fetal death	7	20.0
Respiratory tract infection	7	20.0
Spontaneous preterm delivery	6	17.1
Acute pyelonephritis	2	5.7

**Some patients had multiple complications.*

Table2: *Intrapartum complications in sickle cell disease patients

Complication	†N=31	%
Anaemia	18	58.1
Vasooclusive crisis	4	12.9
Fetal distress	2	6.5
Failure to progress due to CPD‡	1	3.2
None	8	25.8

**some patients had multiple complications.*
† 7 patients had C/S before the onset of labour
‡ CPD=cephalo pelvic disproportion

Table3: *Puerperal complications of patients with sickle cell disease in Pregnancy

Complication	Freq (N=38)	%
Anaemia	22	57.9
Vaso oclusivecrisis	4	10.5
Wound infection(C/S)	2	5.3
Maternal death	2	5.3
Puerperal psychosis	1	2.6
None	11	28.9

**some patients had multiple complications.*

DISCUSSION

This review indicates that pregnancy in sickle cell disease patients is still associated with high maternal morbidity and mortality and poor perinatal outcome in Jos, North Central Nigeria. This may be partly related to the finding in this review which shows that a high proportion of the women booked for antenatal care in the second trimester, indicating that these patients may not

have realized that there pregnancy was associated with higher risk than their counterparts with normal haemoglobin. This is despite the fact that most of them (79%) had more than primary education. This relatively late booking may have accounted for the high incidence of antenatal admissions in this study. It is also worthy to note that one of the patients that died had her antenatal care at a primary health care centre and was only referred in labour.This calls for a need to intensify health education for these patients. Our community health workers also need some education on the risk associated with pregnancy in sickle cell disease patients.

The high incidence of anaemia (62.9%) observed in this study was similar to the finding of a similar study in Ibadan, south western Nigeria[15]. Another similar study in Abraka Delta state Nigeria however observe a higher incidence of 92.25% [11].This study however observed a higher incidence of severe anaemia compared to that observed in Ibadan[15] and consequently, a high frequency of antenatal blood transfusions. This has serious implication for the risk of transmission of HIV and other related infections in these patients.

The chronic haemolytic state of sickle cell disease patients associated with increased requirement for haemopoeitic nutrients in pregnancy increased their susceptibility to anaemia. Prophylactic blood transfusion was suggested in the past as a way of improving the maternal and perinatal outcome;[16] however, randomized studies have not shown any benefit over selective blood transfusion,[17] hence the practice in our hospital has been to transfuse these patients only when clinically indicated.

This study however observed a higher incidence of bone pain crisis (51.4%) compared to those in Abraka, Delta state (23.81%) [11] and Ibadan (7.3%) [15]. This may reflect a true higher incidence of bone pain crisis in this part of the country compared to those of Ibadan and Abraka. However,studies in the United States and nearby Cotonou, Benin republic recorded similar incidences of 50% and 57% respectively of bone pain crisis in larger series[12,13].
Malaria in pregnancy was also a common complication in pregnancy in this series occurring in 25.7% of pregnancies. This was similar to the incidence (22.4%) observed in Lagos[14.] The incidence of pregnancy induced hypertension of 25.7% observed in this study was higher than that observed in Ibadan [15]. The high proportion of nullipara in this study may be responsible for this. Other studies from Atlanta (USA), Jamaica and Baharain showed no significant difference in risk of pregnancy induced hypertension or preeclampsia between patients with sickle cell disease and those with normal haemoglobin genotypes[9, 18, 19]. These discrepancy calls for further research in this area.

Other maternal complications observed included urinary and respiratory tract infections, and pseudotoxaemia, which is consistent with complications recorded in other studies [10-12, 14,15].

There was also a high caesarean section and instrumental delivery rates compared to the institution rates of 17% and 1.4% respectively. Other studies also observed higher caesarean section and instrumental delivery in patients with sickle cell disease in pregnancy [11,15,19]. This could be due to the high incidence of maternal and fetal complications often leading to delivery by caesarean section. Most Obstetricians in our centre routinely shorten the second stage of labour in these patients, this therefore explain the high instrumental delivery rate.

Babies of sickle cell disease patients were also noted to have lower gestational age at delivery and lower birth weights. This is in contrast with the study in Ibadan which observed no difference in gestational age at delivery with patients with normal haemoglobin[15]. It is, however, comparable with findings from studies from Jamaica and in the United States which also observed lower birth weights and gestational age at delivery.[13,18] Factors responsible for lower birth weight for mothers with sickle cell disease are varied and inconsistent, but have included lower gestational age [18], lower maternal weight, history of pre-eclampsia, acute anaemic episodes and the number of sickle cell related complications during pregnancy[10]. Lower gestational age could be influenced by a high frequency of early surgical delivery or premature induction both of which are common interventions in management of pregnancy among mothers with sickle cell disease, though this was not the case in this review..
The high perinatal and maternal mortality observed in this study are in agreement with findings of similar studies within and outside the country [9-11 14,15,18-23].
Though our study is limited by its retrospective nature with only thirty eight of the forty four case files retrieved, the findings are still pertinent.There is need for pre conception care for these patients and closer monitoring during antenatal care, labour and

pueperium by a multidisciplinary team of obstetricians, haematologist and trained sickle nurses.

The importance of early presentation for prenatal care should be stressed to these patients so that antenatal complications can be detected and treated early.

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