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Original article

Sickle cell disease and pregnancy: analysis of 34 patients followed at the Regional Blood Center of Ribeirão Preto, Brazil

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ABSTRACT

Objective: The objective of this study was to verify the evolution of pregnancies in sickle cell patients followed at one institution over a period of 12 years (January 2000 to June 2012).
Methods: The study evaluated 34 pregnant women with sickle cell disease with a mean age of 23.9 ± 5.3 years. The incidence of obstetric complications, non-obstetric complications linked to sickle cell disease and complications in the newborn were analyzed.
Results: A total of 26% of the cases reported previous miscarriages, 20% had preterm labor, 10% had pre-eclampsia, and 5% had gestational diabetes. Forty-one percent of the deliveries were cesarean sections and 29% of patients required blood transfusions. In respect to sickle cell disease, 62% of patients had vaso-occlusive crises, 29% had acute chest syndrome, 23% had urinary tract infection, 15% had impaired cardiac function and 6% developed pulmonary hypertension. Only one patient died in the postnatal period due to acute chest syndrome. The mean gestational age was 37.8 ± 2.63 weeks, and mean newborn weight was 2.809 ± 643.8 g. There were seven fetal losses, including three stillbirths and four miscarriages. The impact of transfusion therapy on the incidence of maternal–fetal complications during pregnancy was evaluated.
Conclusions: Pregnancy in sickle cell patients is still associated with complications. Although no statistical difference was observed between transfused and non-transfused women, there were no deaths (fetal or maternal) in transfused patients whereas one maternal death and three stillbirths occurred in non-transfused women. A larger study of sickle cell pregnant women will be necessary to elucidate the actual role of transfusion during pregnancy in sickle cell disease.

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Introduction

Sickle cell disease (SCD) comprises a group of diseases characterized by the presence of sickle hemoglobin (Hb S). It is classified as sickle cell anemia (Hb SS), hemoglobinopathy SC, hemoglobinopathy SD, S-beta thalassemia (Hb S-beta) and other associations of mutant hemoglobins with Hb S.

In situations of low oxygen tension, Hb S solubility decreases, resulting in the polymerization of these molecules. The intracellular formation of Hb S polymers affects the red cell structure, changing it into a sickle-shaped, thereby damaging the cell membrane, making it more rigid and exposing a greater number of adhesion molecules on the cell surface, thus increasing the adherence of red cells to the vascular endothelium.[1] This phenomenon, named sickling, is responsible for the premature destruction of red cells by the reticuloendothelial system, causing a chronic hemolytic anemia. Under stress situations, such as infections, deoxygenation of Hb molecules and sickling of a large number of red blood cells occur. These cells adhere to the vascular endothelium which may cause vessel occlusion and, consequently, tissue ischemia causes the painful crises that characterize one of the clinical features of this disease. Chronic hemolytic anemia and frequent vaso-occlusive crises cause damage to various organs and impair both the survival and the quality of life of patients with SCD.[2]

Until the 1970s, the management of sickle cell patients was poor and pregnancy was associated with high maternal and fetal mortality.[3] Nowadays, with newborn screening and preventive measures such as vaccination and antibiotic prophylaxis since birth, patient survival has improved.[4] Furthermore, the quality of obstetric and neonatal care has also corroborated to a significant reduction in the maternal mortality rate (from 4.1% to 1.7%) and improved fetal survival (from 60 to 80%).[4,5] However, despite all the medical advances in recent decades, pregnancy in sickle cell patients is still associated with many clinical and obstetric complications compared to the general population.[6–8]

The physiological adaptations that occur in the circulatory, hematologic, renal, and pulmonary systems during pregnancy can overburden organs that already have chronic injuries secondary to SCD, increasing the rate of obstetric complications such as eclampsia and pre-eclampsia as well as the complications of the disease, such as worsening of vaso-occlusive crises and acute chest syndrome.[7]

Objective

The aim of this study was to assess the evolution of pregnancy in sickle cell patients followed at one institution, the Faculdade de Medicina de Ribeirão Preto, Universidade de São Paulo (HC-FMRP-USP) in a 12-year period (January 2000 to June 2012), and discuss the impact of blood transfusion on pregnancy.

This study will contribute to the knowledge on the prevalence of maternal and fetal complications occurring in this population and show the impact of therapeutic measures used to control these complications during pregnancy.

Table 1 – Characteristics of the patients according to their sickle cell genotype.

	Hb SS	Hb S/beta^0	Hb SC
Number of patients	24	7	3
Mean age (years)	22.6	22.0	25.0
Previous HU use – *n* (%)	5 (21)	2 (28)	1 (33)
Blood transfusion – *n* (%)	5 (21)	4 (57)	1 (33)
Obstetric complications – *n* (%)	14 (58)	1 (14)	1 (33)
Sickle cell complications – *n* (%)	18 (75)	6 (85)	2 (67)

Methods

This was a retrospective study that aimed at analyzing the evolution of pregnancies in sickle cell patients during the period covered by the study (January 2000 to June 2012).

Study participants

The subjects comprised sickle cell patients followed at the Hospital das Clinicas, Universidade de São Paulo (USP) in Ribeirão Preto. Patient inclusion criteria were having a diagnosis of SCD (Hb SS, Hb S-beta or Hb SC) by hemoglobin electrophoresis and having had one or more pregnancies from January 2000 to June 2012. The patients were then divided in two groups in order to evaluate the impact of blood transfusions on sickle cell complications during pregnancy.

Design

Clinical data was obtained through a review of medical records from the hospital with the confidentiality of information being preserved. The results of laboratory tests were obtained through the online hospital system using the ATHOS program. The clinical and laboratory data were recorded on a data collection form and later compiled for statistical analysis of the prevalence of maternal and fetal complications.

Statistical analysis

Data are presented as descriptive statistics including means and percentages. The Mann–Whitney non-parametric statistical test was used to compare the transfused and non-transfused groups as the samples did not have a Gaussian (normal) distribution.

Results

The study evaluated 34 pregnant women with SCD; 24 (70.5%) had Hb SS, seven (20.5%) Hb Sb^0-thalassemia and three (8.8%) Hb SC. The mean age was 23.9 ± 5.3 years and 20 (59%) were followed from the first trimester of pregnancy in the High-risk Pregnancy Outpatient Service of the hospital, nine (25%) started this follow-up in the last trimester, and five (15%) did not have any follow-up in this service. The characteristics of the patients according to the type of SCD are shown in Table 1.

Hb SS patients had more obstetric complications (three stillbirths, three miscarriages and eight pre-term labors and one maternal death) than the other two genotypes (S/beta^0 with one miscarriage and Hb SC with one pre-term labor). Most

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