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SICKLE CELL DISEASE AND PREGNANCY: AN INTEGRATIVE REVIEW

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ABSTRACT

Objective: To analyze the scientific productions about gestation in women with sickle cell anemia.
Method: It is an integrative review study carried out in the Virtual Health Library (VHL) database, using the descriptors pregnancy and sickle cell anemia, from 2008 to 2017.
Results: The publications focused on the Southeast region of Brazil in which the number of cases is lower than in the other regions. The complicating gestational factors in sickle cell anemia (HF) were anemia, infections, preterm labor, teenage pregnancy, among others. And factors that increased maternal and fetal risks were spontaneous abortion, early rupture of membranes, restricted intrauterine growth, postpartum infection, among others.
Conclusion: Pregnant women with sickle cell anemia must be accompanied by a multi-professional reference group in high-risk pregnancy, the hematologist and the primary care team, aiming at integral care and the reduction and / or control of maternal and fetal risks.

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INTRODUCTION

Sickle-cell diseases are the most common hereditary diseases in the world. These diseases result from changes in normal hemoglobin (HbA), generating a mutant hemoglobin (HbAS). This mutant hemoglobin (HbS) in pairs with other mutant hemoglobins such as C, D, E, S then constitute the sickle cell group. Because they cause changes in hemoglobin, these diseases are also known as hemoglobinopathies, and despite the variability of severity, they present similar clinical manifestations. Among hemoglobinopathies, sickle cell anemia is the one with the greatest clinical significance and the highest occurrence in Brazil (Paz, 2015). This hemoglobinopathy is present in 5% of the world population in the heterozygous form (AS) and there are 300,000 new births each year worldwide in its most severe form homozygous (SS), (Barbosa, Dias, Abreu, 2012). Pregnancy in sickle cell disease often leads to significant maternal-fetal morbidity, however, this evolution can be avoided by adequate prenatal care.

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Adequate family planning is an important factor in the health care of women with sickle-cell disease, since more than one-third of the pregnancies associated with sickle-cell anemia result in spontaneous abortion, fetal death or neonatal death (Brazil, 2008). It is estimated that in Brazil 6 to 10% of the race / color population of black and brown skin is sickly and 2% in the whole population, this proportion is related to the miscegenation of the country and the origin of the disease itself - African Among the regions is concentrated in the North and Northeast with a prevalence of 6 to 10%, while in the South and Southeast 2 to 3%. Bahia is one of the states with the highest incidence of sickle cell anemia and sickle cell trait, with 1: 650 live births with sickle cell anemia and 1:17 live births with sickle cell trait. The World Health Organization estimates 500,000 births of women with sickle-cell anemia each year (Brazil, 2008, 2012). Because sickle cell anemia is a poorly understood disease, people are not aware of the hereditary factors and symptoms presented for screening and diagnosing the disease early. In addition, women who intend to become pregnant need to be screened in family planning in order to facilitate preconception to achieve a pregnancy without extreme complications (Brazil, 2012, 2015). Pregnant women with sickle cell anemia are usually insecure and not

knowing how to act during this phase of life, as most are not prepared to face the clinical dimension of the disease, another problem is if the partner does not have the disease, (Santos *et al*., 2011). In the present study, it is possible to determine the relationship between the disease and its effects on gestation.
Therefore, the present study is justified by the existing need to analyze published productions of women who experience gestation with sickle cell disease in order to contribute to broadening scientific knowledge in the field of health sciences and to collaborate the efficient performance of health professionals, among them nurses. In this way, the objective was to analyze the scientific productions about gestation in women with sickle cell anemia.

MATERIALS AND METHODOLOGY

Integrative review study of methodological approach, includes several types of research, combines data from theoretical biology, incorporate definitions of concepts, review of theorists of evidence and analysis of methodological problems of a particular topic that generates a consistent and comparative panorama of relevant issues (Souza *et al*., 2010).
The integrative review consists of six phases, which outlined this study. The question was elaborated: What is the knowledge available in the Virtual Health Library (VHL) database about gestation in women with Sickle Cell Anemia?
The search for the scientific productions was carried out in the VHL database using the Health Sciences Descriptors with the following combination: pregnancy AND sickle cell anemia in the "title, abstract, subject". For the selection, the following inclusion criteria were used: a) articles published in full text; b) articles published in the period from 2008 to 2017; c) in Portuguese and English; d) with gestation in Sickle Cell Anemia. Thus, the documentary corpus of this work is constituted of six scientific articles. The data were collected by means of an instrument prepared by the authors containing: title, year, authors, objectives, main results and conclusions (Table 1). The qualitative exploratory approach was used to analyze the articles. Discussions of the main findings and the exposition of the integrative review per se were carried out. The data were analyzed and discussed according to Bardin's (1970) content analysis through the following steps: pre-analysis, material exploration and data processing and data interpretation, with three thematic categories emerging: 1) Characteristics of Scientific Productions; 2) Complicating gestational factors of Sickle Cell Anemia; 3) Gestation in women with sickle cell anemia and maternal fetal risks.

RESULTS AND DISCUSSION

A total of 1,144 articles were found in the virtual health library database, after the application of the inclusion criteria and the title, abstract and complete article, six articles were selected, which are part of the corpus of this study. The methodological approaches used in the articles were qualitative and quantitative in the same proportion, where they point out the interest of evaluating both the perceptions of the subjects and the thoughts of different authors, as well as the way of approaching the problematization. The articles were published in the states of Rio de Janeiro, São Paulo and Minas Gerais in health journals written by health professionals. The Southeast Region is one of the regions with the lowest prevalence rates of sickle cell anemia and the most published in the last 10 years, so it is necessary to pay more attention to the researchers in the Northeast and North regions who presented few publications, respectively and are the regions with the highest number of cases. The studies point to several gestational factors as a consequence of complications of gestation with sickle cell disease, such as maternal mortality, anemia, infections and premature labor. Gestation in women with sickle-cell anemia becomes an aggravating potential for women's health and the concept and can make them worried and insecure (Cox and Beauquier-Macotta, Santos *et al*., 2014). Even so, they dream of fulfilling the desire for maternity, and multi-professional assistance is needed during the period of childbirth and puerperium, a time of great happiness but crucial for the risk of death or of being born with sickle cell anemia (Xavier, *et al*., 2013).

Since the 1972s, maternal and perinatal mortality rates among pregnant women with sickle cell anemia have been very high. Previously, information was provided for women with this pathology to refrain from getting pregnant or to perform tubal ligation, even before pregnancy or after pregnancy and abortion. With advances in medicine since the 1980s, significant improvements in care have been observed, helping the prognosis of these women and the ability to thrive (Nomura, *et al*., 2010). Nowadays, the panorama of these diseases, mainly sickle cell anemia, presents another profile. Although pregnancy carries maternal and fetal risks, advancement in medicine has provided better gestational outcomes in women with sickle cell anemia with high-risk prenatal insertion, rational use of blood transfusions, and stem cell transplantation (Nomura, *et al*., 2010, Paz, 2015). There are several factors that may complicate pregnancy in women with sickle cell anemia due to sickle cell disease, such as hypoxia, changes in the ionic content of red blood cells, dehydration, and intracellular hemoglobin concentration (Silva-Pinto *et al*. al., 2014). Acute thoracic syndrome, infections, severe anemia, cholecystitis, hypersplenism, among others, are intercurrences that need to be carefully evaluated, detected and treated, one of the susceptibilities is premature labor affecting approximately 30% to 50% of pregnancies before of 36 weeks, the average being usually 34 weeks of pregnancy, where this birth may be natural or cesarean according to the need of the patient.

The painful crisis is the most common cause of morbidity among other factors such as pre- and postpartum infection, especially pyelonephritis and pneumonia, anemia, fetal growth restriction, premature birth, stillbirth, spontaneous abortion, worsening of the lesions osseous and retinopathy, low birth weight and pre-eclampsia. In the third trimester of gestation, it is very common that these complications occur, both for pregnant women with sickle cell anemia and for those with sickle cell trait (Nomura, *et al*., 2010; Silva-Pinto, *et al*., 2014). However, these disorders often occur in the case of pregnancy with adolescence, requiring special care due to the early destruction of erythrocytes by the spleen, and the bone marrow cannot replace these cells causing chronic hemolytic anemia. Recurrent vascular occlusion episodes cause discomfort to the mother and restriction of fetal development (Barbosa, Dias, Abreu, 2012). The red blood cells with their sickle shape cause vasoconstriction and decrease the conduction of oxygen and nutrients to the placenta, failing to meet the needs of the concept, exposing the pregnant woman with sickle cell anemia to the risk of abortion and neonatal death (Barbosa, Dias, Abreu, 2012; Silva-Pinto, *et al*., 2014).
In this case, the prenatal care should have specific observations for the early detection of aggravating factors besides the care

Table 1. Documentary Corpus about Gestation in Women with Sickle Cell Anemia, 2008 to 2017

Title	Reference	Objective	Main results	Conclusion
Maternal and perinatal outcomes in gestations complicated by sickle cell disease.	Nomura, *et al*., 2010.	To evaluate the maternal and perinatal outcomes of gestations complicated by sickle-cell diseases, comparing them with sickle-cell carriers.	The complications observed were hospitalization before delivery, occurrence of urinary infection, pneumonia, pulmonary hypertension and restriction of fetal growth	Pregnancy imposes important changes in the body, taking into account the greater incidence of complications that need greater care to minimize or combat them.
Pregnancy experiences reported by women with sickle cell anemia in a university hospital: A qualitative study.	Santos, *et al*., 2011.	To discuss, in the qualitative research approach, the meanings of experiences associated with the particular phenomenon of pregnancy, as reported by women with Sickle Cell Anemia attended at a university outpatient service.	1) Ambiguity between desires and fears about pregnancy: balance / imbalance 2)Lossesand disappointments 3) Emotional meanings of support from family members and staff	Despite the experiences of anguish that normally involve the pregnancy of women with sickle cell anemia, their clinical and emotional conditions did not represent impediments to the clear manifestation of the desire for motherhood.
Pregnancy in adolescence and its interaction with sickle cell anemia	Barbosa, Dias e Abreu, 2012.	To report maternal complications in adolescent pregnancy with sickle cell anemia.	Among the three trimesters of gestation, it was noticed that the pregnant woman underwent blood transfusion, and there was no improvement in the anemia, but the fetus remained healthy. The transfusions performed on the patient during the gestational period occurred to improve the anemia, however, the accumulation of iron in the organism raised the results of ferritin and serum iron.	Coping with the situation of teenage pregnancy and sickle cell anemia is important and should be explored in order for adolescents and their families to understand how to act in their need to reduce fetal and maternal mortality.
Perception of women suffering from sickle cell anemia regarding pregnancy: an exploratory study	Xavier, *et al*., 2013.	To analyze the perception of women with sickle cell anemia in pregnancy.	From the moment the woman is aware of her pathology, she begins to understand the risks that the complications of the disease can cause during the gestational period.	The study points to the lack of information about clarifications for this population, preconception planning and the perception of self care for a peaceful pregnancy.
Maternal representations during a pathological pregnancy: the case of sickle cell anemia	Cox e Beauquier-Macotta, 2014.	To approach representations in the context of pathological pregnancy, in the specific case of sickle cell anemia.	During the study, the number of questions about pregnancy in women with sickle cell anemia was questioned, and the woman questioned her ability to care for and love the child.	It is noticed that mothers with sickle-cell anemia have many concerns about the underdeveloped gestation if their baby will inherit the disease if she will be willing to practice motherhood.
Sickle cell disease and pregnancy: analysis of 34 patients followed at the Regional Blood Center of Ribeirão Preto, Brazil	Silva-Pinto, *et al*., 2014.	To verify the evolution of gestations in sickle cell patients.	The complications observed were hospitalization before delivery, occurrence of urinary infection, pneumonia, pre-eclampsia, gestational diabetes, spontaneous abortion and restriction of fetal growth.	Pregnancy in patients with sickle cell disease 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 stillborn and three stillbirths occurred in non-transfused women.

with vitamin supplementation, guidance on water intake, avoid exposure to sun and cold, and exhaustive physical activities to avoid risk

Conclusion

The scientific productions about gestation in women with sickle cell anemia still deserve attention on the part of the researchers, because it is a pathology that determines complicating factors during gestation, generating risks for pregnant women and for the fetus. It was noticed that, even with the progress achieved in the health area, there are still incipient publications about PA in pregnant women and the safe and protective care so that women with sickle cell anemia can become pregnant without fears and yearnings of the complications that may occur in this period. Thus, it is necessary for pregnant women with sickle cell anemia to be accompanied by a multi-professional reference group in high-risk pregnancy, the hematologist and the primary care team, aiming at integral care and reduction and / or control of maternal and fetal risks. Therefore, it is proposed future investments in studies in this area of high-risk gestation in order to identify the association between facilities for access to specialized health services and the occurrence of maternal and fetal complications during pregnancy in women with sickle cell anemia. And with this help to enable health managers to evaluate the actions established for this population.

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