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PHARMACEUTICAL CARE IN SICKLE CELL DISEASE: LITERARY REVIEW

ATENÇÃO FARMACÊUTICA NA DOENÇA FALCIFORME: REVISÃO LITERÁRIA

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Abstract
The aim of this study is a literary review and to analyze the clinical aspects of sickle cell anemia with the attention of the pharmacist in relation to this disease. A literature review was carried out using Pubmed, Lilacs, Scielo, and Virtual Health Library (VHL) databases. Using texts available in Health Education, which is an essential tool to make patients aware of the characteristics of the disease and promote the correct use of medicines. The research is justified on the role of pharmaceutical care in Sickle Cell Disease in general. The results of this study can serve as support to the literature or even to Organs competent bodies of hospital sectors for implementations and improvements in health.

Keywords: Pharmacist. Sickle Cell Disease. Health Education.

Resumo
O objetivo deste estudo é uma revisão literária a fim de analisar os aspectos clínicos da anemia falciforme com atenção do farmacêutico em relação a essa doença. Foi realizada uma revisão bibliográfica, através das bases de dados Pubmed, Lilacs, Scielo, e Biblioteca Virtual em Saúde (BVS), utilizando textos que dispunham na Educação Sanitária que é uma ferramenta essencial para conscientizar os pacientes a respeito das características da doença e da promoção do uso correto de medicamentos. A pesquisa justifica-se sobre a atuação da atenção farmacêutica na Doença Falciforme de forma geral. Os resultados deste estudo poderão servir como suporte às literaturas ou até mesmo aos órgãos competentes de setores hospitalares para implementações e melhorias na saúde.

Palavras-chaves: *Farmacêutico. Doença Falciforme. Educação Sanitária.*

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Introduction

Sickle cell disease is a type of genetic and hereditary disease. Alteration in the genetic code can result in defective production of the protein hemoglobin, which is responsible for binding and transporting oxygenation. The result of protein alteration is that erythrocytes undergo important changes throughout their morphology, producing sickle-like abnormal sickle cell erythrocytes (GUIMARÃES; COELHO, 2010; HOFFBRAND; MOSS, 2013). Patients with sickle cell have several clinical manifestations and in the most severe forms they can lead to bone marrow necrosis, splenic problems, acute chest syndrome and hemolysis (BRUNETTA et al, 2010).

Diagnosis is performed in the first months of life through isoelectric focusing electrophoresis or high-performance liquid chromatography. The treatment is done by drugs whose most used are Hydroxyurea , Phenoxemethylpenicillin , Potassium, Benzylpenicillin , Benzathine and Erythromycin Stolate (BRASIL, 2016).

Sickle cell diseases are the most prevalent hereditary diseases in the world, with the highest prevalence of S (HbS) in tropical Africa (BRAZIL, 2016). In Brazil, it is considered a health problem with more incidents in the states of Bahia, Rio de Janeiro and Minas Gerais (CAVALCANTI; MAIO, 2011). According to studies, in Brazil, there are about 25 to 30 thousand carriers of the disease (CANÇADO et al, 2009), affecting around 0.1 to 0.3% of the Brazilian black population (LOUREIRO; ROZENFELD, 2005). It is estimated that around 3,500 new cases are diagnosed annually (CANÇADO et al, 2009).

Given the severity and destruction of sickle cell disease, early diagnosis and treatment of this disease has a great impact on the quality of life of patients. Aiming at this, the pharmacist plays an important role in genetic counseling, diagnosis and patient treatment, in which pharmaceutical care and care becomes an effective and necessary tool. Thus, this work aims to demonstrate possible treatments, symptoms and the importance of the pharmacist in sickle cell disease through a retrospective study and related publications.

Pharmaceutical attention

In 1990, “ Hepler and Strand used for the first time in the scientific literature the *term* Phamarceutical *Care ”,* which has been translated in our country to Pharmaceutical Care. Therefore, pharmaceutical care is the responsible provision of pharmacological treatment with the aim of achieving satisfactory health outcomes, improving the patient's style and quality of life. Pharmaceutical care is a sequence of steps of clinical methods. The clinical method includes data collection, problem identification and implementation of a plan for patient care and follow-up.

sickle cell disease

Sickle cell disease is a genetic alteration, characterized by a type of mutant hemoglobin called hemoglobin S (or Hb , S) that causes the erythrocytes to distort, making them take the shape of a “sickle” or “half-moon”. Sickle cell disease is a genetic

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issue, that is, inherited from parents to children. The parents are always carriers of S or C trait or heterozygotes or beta thalassemia or have sickle cell disease.

Sickle cell anemia (SCA) meets these conditions and involves psychological and social aspects that are difficult to manage, which is why patients with this disease need special care and comprehensive care. The disease has been studied from a molecular and epidemiological point of view, but there is little information on special care for this type of patient, while information on medication care is zero. (PAIVA and SILVA, RAMALHO & CASSORLA, 1993). Sickle cell anemia is the most common inherited blood disorder in the world. It is estimated that 1: 7,400 people born in the state of São Paulo are affected each year (MAGALHÃES et al., 2009).

Complications of the Disease

Pain

An important aspect, especially in the care of these patients, is adequate pain management. The pain is due to the difficulty in blood circulation, which has increased viscosity due to sickled red blood cells, which can cause small vessels to clog. Denser cells, with higher concentrations of Hbs , have a lower affinity for oxygen and high viscosity, and may lose their deforming power, which hinders circulation through small-caliber vessels (FIGUEIREDO, 2007). Pains located in the hands and feet, and called dactylitis and very common in children up to five years of age (ANVISA, 2002).

Jaundice

When bilirubin levels are high in sickle cell patients, it generates jaundice, resulting from the accelerated destruction of red blood cells, generating an excess of their precursors and a precipitated formation of bile salts. Jaundice is a yellowing of the eyes and skin due to the accumulation of pigment in the tissues. The worsening of jaundice can lead to cholelithiasis, with multiple and pigmented stones, requiring surgical intervention in most cases (cholecystectomy) (GUMIERO et al., 2007).

infections

cell patients suffer from functional asplenia that causes immunosuppression. This occurs because the spleen is an organ that produces defense cells such as B lymphocytes, which synthesize antibodies against pneumococci and influenza type B (Hib) hemophilia. Pneumococci are considered the main agents related to those obtained associated with infections. (LOGGETTO et al, 1999). An infection is accompanied by dehydration and low blood oxygen tension, the painful crises become stronger, taking into account that the molecules increase the adhesion of red blood cells to the endothelium of the blood vessel facilitating vaso-occlusion. (DI NUZZO and FONSECA, 2004).

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Brain stroke

Stroke occurs due to lesions that can be ischemic or hemorrhagic. In ischemia, blood flow is blocked by lack of oxygen in certain parts of the brain, leading to changes that may or may not express clinical symptoms. Bleeding includes blood vessel overflowing. In sickle cell patients, silent effusions are more of an event, visual changes and attention and memory deficits (PLUMACHER et al., 2004).

Acute splenic sequestration crisis

Acute splenic sequestration crises (ASCS) are the major causes of mortality among sickle cell disease patients, occurring in up to 30% of these patients before the age of five years. It is characterized by the enlargement of the spleen, resulting from the accumulation of red blood cells within the organ, with a consequence of drops in circulating levels of red cells in peripheral blood of at least 2g/dl of the patient's baseline values (ANVISA, 2002). Because of this, patients are at risk of presenting hypovolemic shock, and may die within hours. The most effective way to reverse this condition of ACS is to perform a transfusion of red cells, since the detection of the episode is early and it may be necessary to perform a splenectomy operation (CANÇADO et al, 2009).

Some authors indicate a relationship between the occurrence of CSEA and infections, but this could not be proven. Noting that patients with higher levels of HbF are less likely to develop ACS (CANÇADO et al, 2009). The most recurrent attacks are in about 50% of patients who survive the first episode, with 20% mortality in these patients (ANVISA, 2002).

Acute chest syndrome

Vascular occlusion is common in the lungs of sickle cell patients, with water chest syndrome (WAS) being the main cause in adults that can lead to death (GUALANDRO, FONSECA & GUALANDRO, 2007). ACS has the characteristic of a series of symptoms that can start with fever and progress to a cough with dyspnea, chest pain and pulmonary infiltration evidenced by radiography (GUALANDRO, FONSECA & GUALANDRO, 2007).

Treatment

Concerning the clinical picture of the sickle cell disease patient, there is no specific treatment. Regarding the treatment, clinical protocol and therapeutic guidelines for sickle cell disease, priority is given to the prevention of vaso-occlusive crises, pain, aplasia, splenic sequestration, thoracic water and neurological with several complications, the prevention of infections in the first days and life, aiming at an improvement in the survival, the quality and well-being of patients. Other procedures are adopted as a means of minimizing its effects, such as: chelating, palliative and preventive drugs. (BRAGA, 2007; BRUNETTA, et al, 2010).

Most crises in sickle cell disease patients are characterized by episodes of severe pain. Some medications can relieve pain. Drugs such as paracetamol are indicated in mild cases, as it is effective in secondary pain. Most patients may have a

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greater number of crises, and when the number is above six episodes of crises for a period of one year, the use of hydroxyurea is indicated, this drug generates a positive result, considering that it stimulates the production of fetal hemoglobin, helping to prevent the acute syndrome (SANTOS, 2009). In addition to hydroxyurea , other drugs can be used, such as Potassium phenoxemethylpenicillin , Benzylpenicillin , Benzathine , Erythromycin stolate (BRASIL, 2016). Hydroxyurea is a drug that has shown many positive results regarding the treatment of sickle cell disease, which began to be part of a therapeutic set for patients in 1998 and since then has been shown to be effective in preventing clinical complications by improving quality of life (CANÇADO et al, 2009).

MATERIALS AND METHODS

The proposal developed in this study was to understand pharmaceutical care in sickle cell disease and some possible pharmacological treatments. The research was carried out through literature review and theses and articles published in Google Scholar and Scientific Electronic Library Online (SciELO). A bibliographic research was carried out using the following descriptors: Sickle Cell Disease, Pharmaceutical Care, Health Education. Full articles related to the topic published between 1990 and 2020 were used in the study. All scientific productions that did not meet the criteria described above were excluded.

RESULTS AND DISCUSSION

The role of the pharmacist in sickle cell disease

The important role of the pharmacist in pharmaceutical care, acting to guarantee access to the drug and its rational use. “According to Novaes (2009) the performance of therapy and diagnosis of patients, in many cases, will generally depend on the care provided by a multidisciplinary team.” Thus, it is important to reinforce good integration and communication with professionals and participants of the entire team so that the patient is well attended. We can discard the importance of the Pharmacist within the multiprofessional team, as they are responsible for all pharmaceutical care to ensure the correct and safe use of medicines.

Pharmaceutical care has great importance in returning and carrying out activities that cover the correct and rational use of medicines. With this relationship, the pharmacist is able to identify and correct certain problems that may be related to medical prescriptions such as: drug interactions, drugs with the same therapeutic indication, drugs without dose, drugs with altered dosage and drugs without a route of administration and drugs that are out of date. of standardization (REIS, 2013). Sometimes patients with sickle cell disease do not have many resources or do not find what they are looking for in the health system. In some certain situations the pharmacist will be the last to have contact with the patient and will have as a health professional, before using any medications. In this way, pharmaceutical care is of great importance and being prepared to be able to serve well and have adequate knowledge

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to offer the patient. (PEREIRA; NUNS, 2008). In addition, pharmacists work in neonatal screening and offer this genetic counseling, thus guaranteeing diagnosis of quality and care. (REIS, 2013) .

CONCLUSION

Therefore, through these studies, it is concluded that the role of the pharmacist in sickle cell disease is essential for carrying out activities and well-being, which go beyond some standard functions such as guidance, monitoring of drug therapy and the best quality of life for the patient. . The pharmaceutical professional is indispensable in the positive evolution of the sickle cell patient with a reduction in the occurrence of complications.

Pharmaceutical care is increasingly inserted in health services, its performance is beneficial both from the clinical point of view and from the economic point of view, improving and also contributing to a better quality therapy for patients and with the support of the prescriber, aiming at o mainly the rational use of medicines.

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