

Editorial

Guidance for assisting patients with sickle cell disease to reduce the impact of the Covid-19 pandemic

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EDITORIAL

Severe acute respiratory syndrome coronavirus 2, also known as COVID-19, has spread to more than 213 countries with almost 12,6 million cases as July 2020 since first reported [1-3]. The clinical features of this disease are not completely understood, however severe illness is thought to predominantly occur in adults with advanced age and those with underlying comorbidities [4]. Sickle cell disease (SCD), an immuno compromised condition, puts patients at higher risk for respiratory infections and subsequent pulmonary complications such as acute chest syndrome (ACS) [5]. In addition, they can evolve with damage to target organs over the years, with the development of heart, lung, neurological and kidney diseases [6].

Some reports suggest that painful vaso-occlusive crisis (VOC) and/or ACS could be triggered by COVID-19 [7], and others suggest that the association between has a high risk for a severe disease course and a high case-fatality rate [8]. Although it is necessary to wait for more experience in relation to the clinical presentation of COVID-19 in SCD, these important points need to be taken into account [6,9,10].

The experience in hematology and blood transfusion, which transforms administration practices excessively focused on procedures, norms and strategy into approaches based on results that determine the citizens' satisfaction, presents a challenge. The great difficulty in the quality of administration of a public service is to obtain effective results by decreasing or eliminating problems, or by adding benefits and values expected by society. The Fundação Hemominas a public health institution, reference in the treatment and follow-up of patients with SCD, aware of the importance of its role as a service provider in a critical and vital area, elaborated strategies to meet a management system based

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on constitutional and institutional public values and quality principles with defined goals accompanied by a appraisal system.

The Hemominas works as a network, with administration and technical coordination located in Belo Horizonte, Minas Gerais (Brazil). It has 23 regional units throughout the state of Minas Gerais, Brazil. Considering these facts, the Fundação Hemominas [11], developed this guidelines:

1. Social isolation: the patients should avoid contact with people with respiratory symptoms or fever
2. The patients should prioritize self-healing, wash their hands and hydrate themselves frequently
3. Patients should seek emergency medical services if they experience fever and symptoms respiratory (difficulty in breathing, persistent cough, nasal wing bleeding).
4. Patients with SCD and respiratory symptoms or VOC should have a chest X-ray [12].
5. Must remember that Acute Chest Syndrome (ACS), a serious complication of sickle cell disease, maybe triggered by respiratory infections, including COVID-19. In addition, infection by coronavirus can lead to hypoxemia, dehydration and metabolic acidosis, triggering VOC including ACS. Therefore, doctors should recognize signs of rapidly progressive ACS, including multi-organ failure, hepatic dysfunction, thrombocytopenia and acute kidney injury [13]. Patients with ACS should be hospitalized for intensive care¹².
6. Maintain control medications such as folic acid, penicillin V, hydroxyurea and analgesics.

7. In SCD patients diagnosed with COVID-19, manifesting fever and cough, have worsening anemia, evidence of hypoxia and/or lung imaging changes, there is a recommendation for early aggressive or exchange blood transfusions [13,14].
 8. Considering pediatric patients with SCD, for those receiving regular blood transfusion therapy for primary or secondary stroke prevention, in areas that may have a deficiency in blood supply, a low dose of hydroxyurea is recommended [12,13,15].
 9. In case of doubts regarding the hematological patients characteristics, the Fundação Hemominas is available to discuss cases through a call center called LigMinas 24 hours a day.
 10. In addition to care, it is important to cooperate with the scientific community, for this, doctors may report cases of COVID in people with SCD to the website <https://covid sickle cell.org/>, only confirmed cases and report after sufficient time has passed to observe the disease course through resolution of acute illness and/or death. The purpose is to identify the impact of COVID-19 on patients with SCD and understand the impact of risk factors such as genotype, prior complications, or SCD related medications, on health outcomes [15].
- The Fundação Hemominas seeks, through these strategic actions implemented during the pandemic, to minimize the impact of Covid 19 on the well-being of patients with sickle cell disease.
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