ISSN: 2573 - 9611

Short Communication

Journal of Pediatrics & Neonatal Biology

World Sickle Cell Day to Call for Maximal Protection of African Sicklers During the COVID-19 Pandemic

Georges Pius Kamsu Moyo*, Laura Kuate, Daniel Armand Kago Tague, Annick Tchouamou, Félicitée Nguefack, Evelyn Mah, Andreas Chiabi

Faculty of Medicine and Biomedical Sciences, The University of Yaoundé I, Yaoundé, Cameroon.

***Corresponding author**
Georges Pius Kamsu Moyo, Faculty of Medicine and Biomedical Sciences, University of Yaoundé. Cameroon.

Submitted: 15 Nov 2020; **Accepted**: 20 Nov 2020; **Published**: 17 Dec 2020

Abstract

Sickle Cell Disease (SCD) is a chronic disease affecting multiple organ systems, causing progressive impairment and loss of function. The COVID-19 infection on its part is a morbid state which could be fatal, especially among populations at risk. The SCD-COVID-19 comorbidity is feared, as it is susceptible to induce the Acute Chest Syndrome, which is a deadly condition in sickle cell patients. There is enough concern about the possible interaction of these two conditions during the ongoing COVID-19 pandemic, especially in Africa and Sub-Saharan Africa where the greatest prevalence of SCD is recorded. This year, the World Sickle Cell Day served as a golden opportunity to stress for a maximal protection of sickle cell patients in Cameroon in particular and Africa in general. Given that the limited health resources in our context may be quite disadvantageous for the effective management of such pathologies with poor prognosis, it appears that primary prevention may be the best option left. This could be done through the strict observation of physical distancing, barrier measures and effective usual prophylaxis in sickle cell patients. More so, the prompt implementation of public health policies aimed at preserving vulnerable groups including sickle cell patients may as well contribute for a maximal protection of such sub-populations at risk.

Key words: COVID-19, Sickle Cell Disease, Africa

Background

The World Sickle Cell Day was established by the United Nations' General Assembly on December 22nd 2008 in order to increase the awareness about the Sickle Cell Disease (SCD) and its treatment among the general public [1]. It was celebrated for the first time on June 19th 2009 and from then, is commemorated every year in all countries on the same date. The 2020 World Sickle Cell day occurred in a particular context, when the world is going through one of its most devastating and deadliest recorded critical phenomenon, known as the COVID-19 pandemic.

In effect, by mid April 2020, the COVID-19 infection which started in early December 2019 in Wuhan, Hubei province in China had spread to more than 184 countries, with over 1.5 million diagnosed cases in all continents [2, 3, 4]. By the same period 10759 cases had been confirmed in 45 countries in Africa with 520 deaths, among which 730 cases and about 10 deaths in Cameroon [5, 6].

Modelling studies show different clinical courses of the pandemic across continents, countries, populations, communities and even age groups [7]. A milder progression of the disease is noted in Africa and is even less in sub-Saharan Africa, probably because of a heavy demographic imbalance, characterized by a considerable shift towards a younger population with less comorbidities [8]. Whereas higher incidences are reported in subjects beyond 50 years of age [9]. However, there is need for vigilance in Africans as statistics from the USA reveal black Americans of African descent are disproportionately more affected than other ancestries [10].

Although a number of research studies seem to corroborate about the fact that the pediatric subpopulation may be the least affected of all, the incidence and the severity of the infection is however determined by the presence of comorbidities including pre-existing conditions such as diabetes, HIV, liver, renal and cardiovascular conditions, chronic obstructive pulmonary disease and immunosuppression [11].

In some patients, these comorbidities may be caused or be associated with Sickle Cell Disease (SCD). As a matter of fact, it is estimated that approximately 5% of the world's population carries

trait genes for hemoglobin disorders of which SCD is the most predominant [11, 12]. SCD affects more than 100000 people in the USA, but over 75% of all sickle cell patients are found in sub-Saharan Africa, with 40% occurring in central Africa [11, 12]. As of now, the disease could still be considered as neglected in the tropics because of the limited number of specialist and the consequent high under-5 mortality rate associated [11]. Despite considerable efforts put in place to improve the management of SCD through early screening, prophylaxis with folic acid and penicillin, immunization and hydroxyurea medication, one of the achievements of current medical sciences is to have partially converted the illness from a "fatal childhood disease" to a "chronic condition of adulthood" [11, 13].

There is significant concern about the comorbidity or overlapping of the COVID-19 infection and SCD with potential triggering of the Acute Chest Syndrome (ACS) which may result in further respiratory complications, thereby worsening the condition of sickle cell patients [14, 15]. This possible interaction between the COVID-19 and SCD should be dread, especially where SCD is prevalent, such as in Africa in general and sub-Saharan Africa in particular. This interaction may even be worse in sickle cell patients with further comorbidities [14, 15]. For these reasons, patients with SCD have been included in the "high risk" group of the population with regards to the COVID-19 pandemic [11, 16, 17]. Moreover, the pre-existing financial burden associated with the management of SCD alone makes diagnostic procedures and management of associated comorbidities such as the COVID-19 unaffordable, especially when no specific subvention is provided to support such patients. Therefore, logistic challenges in meeting the healthcare needs of sickle cell patients during the COVID-19 is to be considered as well [11].

COVID-19 and SCD comorbidity

Acute respiratory illnesses are a major cause of morbidity and mortality in patients with SCD [11, 12, 18]. There is evidence that during infectious respiratory epidemics, children with SCD may be as much as 56 times more affected than others [18]. In effect experience from the 2009 influenza A H1N1 pandemic highlighted an increased susceptibility of children with SCD to develop severe complications due to the respiratory virus, with up to 50% increase in the rate of hospitalisation within this subpopulation [19]. There are few literatures about the vulnerability of sickle cell patients to COVID-19 infection and the effects of the virus on the pre-existing pathophysiological anomalies in sickle cell patients is still being investigated [18]. Though the COVID-19 infection may cause multisystem lesions, it is admitted that the virus is primarily transmitted through respiratory droplets with the respiratory and pulmonary system as main tropism [18]. As a lung infection, the corona virus may alter breathing and respiration causing hypoxia and acidosis, fever and polypnea responsible for dehydration [11, 12, 18]. The hypothesis over a consequent increased rate of sickling and vaso-occlusive crisis causing Acute Chest Syndrome (ACS) has been evoked, as well as suggestions about a more severe viral pneumonia with bacterial superinfection, similar to that of Streptococcus pneumoniae aggravated by hyposplenism [18]. What seems constant is the fact that the frequently affected and particularly vulnerable lungs of sickle cell patients may serve as a breeding ground for severe pneumonia due to COVID-19, with possible secondary bacterial infection which in most cases would cause the ACS. This is thought to be responsible for acute respiratory distress, being fatal in 5% of such patients [18]. The ACS is a term used to describe a number of findings including chest pain, cough, fever, hypoxia and lung infiltrates as may be revealed by chest X-ray [12]. ACS may occur as a result of increased sickling in small blood vessels, pulmonary infarction or emboli, viral or bacterial pneumonia [12]. It is a major cause of morbidity and mortality in SCD, as its management may be quite challenging [12]. Moreover, the use of hydroxyurea in sickle cell patients may be responsible for cytotoxic affects, causing relative immunosuppression which may compromise the outcome of COVID-19 infection in these patients [18].

All these factors make the setting of a particular care pathway for the management of sickle cell patients during the COVID-19 pandemic a necessity, especially knowing that they may be frequent users of emergency departments, which further expose them to infection.

Protecting sickle cell patients from COVID-19

Even though guidelines for the prevention of COVID-19 infection in sickle cell patients have been published by a number of Sickle Cell Disease Associations worldwide, they may not accurately fit to all contexts in our opinion [12, 18]. Taking into account contextual variability and the characteristic limitation of financial and logistic resources prevailing in most African countries, we here suggest adapted specific measures, as well as some preventive public health policies [8].

Specific measures

These are mainly physical distancing, barrier measures, and effective usual prophylaxis including medication which should be rigorously observed by sickle cell patients in order to prevent on one hand the COVID-19 infection and on the other hand, to prevent or improve the home management of pain crisis.

In order to achieve these, regular hand washing with soap or hydro-alcoholic sanitizers is recommended [20]. Patients should use disposable tissues for sneezing, coughing and nosal blowing. Avoid touching eyes, nose, or mouth with unwashed hands. In the absence of disposable tissues, sneezing and coughing may be done into the elbow. One to two meters distancing with people should be observed and the wearing of adequate face masks as from 2 to 3 years of age being systematic. Frequently touched objects and surfaces should be regularly disinfected and contact with people manifesting respiratory symptoms should be avoided. Patients should avoid crowding and stay at home as much as possible [20].

As far as some hygiene and dietary measures are concerned, sickle cell patients are always advised to drink enough water, at least 1.5 to 2l per day in average, they should have a balanced diet, enough sleep with at least 7h per night, avoid high altitudes, unventilated and contaminated atmosphere. Moderate physical exercise is recommended and patients should avoid tight dresses. All routine patients' appointments may be converted to virtual, mail-based or telephonic as much as possible to keep-on following patients while maintaining physical distancing. Patients may be interviewed by telephone, they could transfer their monitoring parameters to their healthcare provider via texting systems, with temperature monitoring reported by phone or presented through video conferenc-

ing [8]. Patients and parents should be educated about COVID-19 signs and symptoms and the importance of physical distancing to limit chances of exposure and infection [8, 18]. Emotional connection with relatives through virtual or internet-based means is encouraged [8]. Patients and parents should continue to seek medical assistance for fever, but should always notify their healthcare providers for guidance where to get safe assessment and management [12]. All patients should have and know how to use a thermometer. Patients are advised to have sufficient supply of all prescribed medication at home, including analgesics to manage both acute and chronic pain [12]. Angiotensin-Converting Enzyme (ACE) Inhibitors and corticosteroids should be avoided as much as possible, given that they may induce ACS. Sickle cell patients on hydroxyurea and other chronic medication should stick to their prescription and make sure they do not lack. The medical staff should encourage patients without fever or signs of infection to manage pain at home with oral medications so as to reduce hospitalizations and visits to the emergency department (ED).

Patients suspected for COVID-19 symptoms such as fever, cough, or shortness of breath should be scheduled for outpatient immediate visit. The emergency department should be avoided as much as possible [12, 20]. If the ED must be used, patients should call ahead to facilitate care and isolation, with triage appropriately done by the health staff, discriminating, so as to protect sickle cell patients when need be [20]. The suspected patients may be tested for COVID-19 in the health center or appropriate sample may be collected and sent to a testing facility for outpatients. Nevertheless, the standard care for managing SCD and fever including culturing of blood and other specimen as indicated should be maintained, just as the administration of empiric broad-spectrum antibiotics to cover encapsulated bacteria. In all cases, assessing patients regularly for signs of acute chest syndrome should be done [12].

For sickle cell patients whom are positive for COVID-19, standard national treatment protocols should be applied as recommended by authorized societies. In Cameroon for instance, the Society of Pediatrics, just as the Society Intensivists recommend hydroxychloroquine, macrolides, zinc and vitamin C supplementation should be administered in first intension, with closer monitoring for signs of ACS and rapidly initiating the treatment if need be [20]. This could include additional empiric antibiotics such as third generation Cephalosporine and aminoglycosides. Supplemental oxygen, incentive spirometry, and good pain control to reduce atelectasis should be envisaged as well. Blood transfusion is performed in patients with worsening anemia, hemoglobin < 9 g/dl or a greater than 2 g/dl fall from basal hemoglobin with evidence of hypoxia or other signs of decompensation, and chest x-ray changes [20, 12].

Public health policies

Given that an outbreak of COVID-19 in regions with high SCD prevalence may be deleterious, with the possibility of overwhelming the actually limited health resources, there may be need for anticipation and preparedness from a public health perspective. Sickle cell patients as other high risk subpopulations should be considered in the development of preventive healthcare strategies [11]. However, various interventions would be more effective only after the size of the population at risk is estimated through large scale screening [21].

The ongoing pandemic may serve most African countries as an opportunity for mobilizing resources to improve health services at large, and specific centers, departments or wards for the management of sickle cell patients [11]. Provisions for the management of SCD and associated comorbidities including the COVID-19 may be permanently adopted as a public health program or grafted to the national program for the fight against epidemics.

Furthermore, a greater decentralization of resources may enable proximity in the care for sickle cell patients, as near as in primary health facilities. Mass communication and education of the public about the COVID-19 risk in sickle cell patients would further increase awareness and solidarity. Protection material such as face masks, hand sanitizer and soap should be provided for such patients in order to reduce financial burden. The epidemio-clinical association between SCD, malaria, various viral and bacterial infections and their prevalence in developing countries allows suggestions for including SCD among targets for the Integrated Management of Childhood Illnesses (IMCI) [11]. It may equally be suggested that sickle cell patients be prioritized when a preventive or therapeutic vaccine against the COVID-19 is available, and possibly introduced among routine or recommended vaccines for sickle cell patients.

Conclusion

The SCD and COVID-19 comorbidity is dread, as it is susceptible to induce complications in sickle cell patients, thereby worsening their prognosis, especially from the respiratory stand point. Primary prevention appears to be the best option for African sicklers given the characteristic limited health resources prevailing in our context. This may be achieved through physical distancing, barrier measures, effective usual prophylaxis in sickle cell patients and the implementation of public health policies aimed at preserving vulnerable groups.

Acknowledgements

All collaborators to this project

Funding

Private

Conflict of Interest

The authors declare that they have no competing interest.

References

1. Serjent GR (2017) World Sickle Cell Day: lessons for India. Indian J Med Res. Jun;145: 705-707
2. Qun Li, M Med, Xuhua Guan, Peng Wu, Xiaoye Wang, et al. (2020) Early Transmission Dynamics in Wuhan, China, of Novel Coronavirus–Infected Pneumonia coronavirus–infected pneumonia. N Engl J Med. 382: 1199-1207.
3. Huang C, Wang Y, Li X, et al. (2020) Clinical features of patients with 2019 novel coronavirus in Wuhan, China. Lancet. 395: 497-506.
4. https://coronavirus.jhu.edu/map.html Last accessed on April 9, 2020.

5. WHO (2020) COVID-19 situation update for the WHO African Region—Sitrep 07 April 2020. April 15, 2020.
6. John Hopkins University Coronavirus Resource Center.
7. Wu Z, McGoogan JM (2020) Characteristics of and important lessons from the coronavirus disease 2019 (COVID-19) outbreak in China: summary of a report of 72 314 cases from the Chinese Center for Disease Control and Prevention. JAMA 2020. 323: 1239-1242.
8. Georges Pius Kamsu Moyo, Dany Hermann Ngwanou, Daniel Armand Tague Kago, Nelly Kamgaing, Ginette Claude Mireille Kalla et al. (2020) Reviewing the Insights of "Confinement" and "Social Distancing" as Measures Involved in the Prevention of the COVID-19 Pandemic. 4: 2020. ABEB. MS.ID.000595.
9. Riou J, Hauser A, Counotte MJ, Althaus CL (2020) Adjusted age-specific case fatality ratio during the COVID-19 epidemic in Hubei, China, January and February 2020. medRxiv 2020; published online March 30. DOI:2020.03.04.20031104 (preprint).
10. Garg S KL, Whitaker M, et al. (2020) Hospitalization Rates and Characteristics of Patients Hospitalized with Laboratory-Confirmed Coronavirus Disease 2019 — COVID-NET, 14 States, March 1–30, 2020. MMWR Morb Mortal Wkly Rep. ePub: 8 April 2020. DOI:http://dx.doi.org/10.15585/mmwr.mm6915e3external icon.
11. Daniel Dexter, David Simons, Charles Kivaga, Nathan Kapata, Francine Ntoumi, Richard Kock et al. (2020) Mitigating the effect of the COVID-19 pandemic on sickle cell disease services in African countries The Lancet Haematology 7: e430-e432, 2020
12. Medical and Research Advisory Committee Sickle Cell Disease Association of America (2020) Sickle Cell Disease and COVID-19: An Outline to Decrease Burden and Minimize Morbidity. March 18, 2020.
13. Ware RE (2013) Is sickle cell anemia a neglected tropical disease? PLoS Negl Trop Dis 7: e2120.
14. Marco Cascella, Michael Rajnik, Arturo Cuomo, Scott C Dulebohn, Raffaela Di Napoli Features, et al. (2020) Evaluation and Treatment Coronavirus (COVID-19). StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2020 Jan. 2020 May 18. PubMed | Google Scholar
15. Faiz A Hussain, Franklin U Njoku, Santosh L Saraf, Robert E Molokie, Victor R Gordeuk, et al. (2020) COVID-19 infection in patients with sickle cell disease. Br J Haematol. Jun; 189: 851-852.
16. Scott D Grosse, Isaac Odame, Hani K Atrash, Djesika D Amendah, Frédéric B Piel, et al. (2011) Thomas N Williams. Sickle cell disease in Africa: a neglected cause of early childhood mortality. Am J Prev Med. 2011 Dec;41: S398-405.
17. Serjeant GR (2013) The Natural History of Sickle Cell Disease. Cold Spring Harb Perspect Med. Oct 1: a011783.
18. Joan-Lluis Vives Corrons, Vincenzo De Sanctis (2020) Rare anaemias, sickle-cell disease and COVID-19. Acta Biomed 91: 216-217.
19. Inusa B, Zuckerman M, Gadong N, et al. (2010) Pandemic influenza a (H1N1) virus infections in children with sickle cell disease. Blood 115: 2329-2330.
20. Cameroon Society of Pediatrics (2020) Recommendations for the Diagnosis and Management of the COVID-19 Infection in Neonates and Children. April 2020.
21. Kiyaga C, Hernandez AG, Ssewanyana I, et al. (2019) Sickle cell screening in Uganda: high burden, human immunodeficiency virus comorbidity, and genetic modifiers. Pediatr Blood Cancer 66: e27807.

Copyright: *©2020 Georges Pius Kamsu Moyo. This is an open-access article distributed under the terms of the Creative Commons Attribution License, which permits unrestricted use, distribution, and reproduction in any medium, provided the original author and source are credited.*