



# Sickle Cell Disease and COVID-19

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## Abstract

**Context:** The COVID-19 pandemic emerged in China and spread to the majority of countries worldwide, causing numerous health problems and restrictions on the people's lifestyles. The complications of this disease can be much more severe in patients who have a weakened immune system or a chronic disease. Sickle cell disease is a genetic disorder in which the immune system is impaired. Exposure to COVID-19 will have severe consequences in the sickle cell disease patients. In this study aimed to investigate the relationship between this disease with the COVID-19 pandemic.

**Evidence Acquisition:** Embase, Scopus, Pub Med databases were reviewed. Keywords were combined as ("COVID19" OR "Coronavirus" AND "Anemia sickle Cell" OR "Sickle cell disease"). The results of related articles were used.

**Results:** At the time of the COVID-19 pandemic, chronic patients and those with weakened immune systems need to be more socially inclined to be less exposed to the COVID-19 virus.

**Conclusions:** When producing the COVID vaccine, high-risk groups such as sickle cell anemia patients should be given priority.

**Keywords:** COVID-19, Coronavirus, Anemia Sickle Cell, Sickle Cell Disease

## 1. Context

In late December 2019 in Wuhan, Hubei Province, China, there came a report on several consecutive cases of acute respiratory syndrome. After health research, the syndrome was diagnosed as zoonosis, and its primary origin was the seafood market. Having been spread to many cities and countries, such as Hong Kong, Taiwan, and Macau and other

continents, this disease, later known as COVID-19, became a pandemic and caused countless health problems in the world (1-5). COVID-19 can be transmitted through respiratory droplets and human carriers (6). The symptoms vary from mild, moderate, and severe. Mild to moderate symptoms appear two weeks after exposure to the virus, including fever and dry cough (7, 8). Severe symptoms include difficulty breathing, which mostly leads to hospitalization in the ICU (9).

About 5% of the world's population carries a gene that disrupts the hemoglobin production chain. The most common disease is sickle cell disease (10). Defects in hemoglobin synthesis cause red blood cells to become crescent-shaped or sickle-shaped. These abnormal conditions are more likely to occur in the absence of oxygen. This lack of oxygen may occur in the face of an infectious agent. The sickle shape of red blood cells causes blockage of arter-

ies and complications in various organs (11-13). One of the high-risk groups of COVID19 is sickle cell anemia (SCD) patients. These patients may be more vulnerable due to immunodeficiency and thrombosis (14).

## 2. Evidence Acquisition

The present study aimed to investigate the relationship between sickle cell disease and COVID-19 disease. To find related articles, three databases, Embase, Scopus, and PubMed were examined. The combination of search keywords is ("Coronavirus" OR "COVID19" OR AND "Anemia sickle Cell" OR "sickle cell disease"). Articles found from databases were collected in EndNote X8 software. Duplicates were detected using this software. The titles and abstracts articles were studied. Then, the full text of the articles was reviewed, if available. Beneficial findings were extracted by studying the articles. was reviewed if available. Beneficial findings were extracted by studying the articles.

## 3. Results

After reviewing the databases, 673 articles were identified. Six articles were duplicates and were deleted. 667 titles and abstracts were studied. Seventy-three full texts

of the article were read. Eight completely related articles were identified, and their information was used. Significant findings were obtained on the relationship between sickle cell anemia and COVID-19 disease.

Fever is one of the main symptoms that is common in both sickle cell disease and COVID-19 disease, so the patients may not be diagnosed if they develop COVID-19 disease. It may be more common in areas of Africa where diagnostic facilities for COVID-19 are more limited.

The decreased effect of serum cytokines is one of the side effects of COVID-19 in some of them (15). Mortality from COVID disease is higher in patients with sickle cell disease and chronic respiratory or cardiovascular disease and hypertension than in patients with sickle cell disease alone (16). Examination of blood cells in patients with COVID-19 and sickle cell anemia revealed that all blood cells were stable, and only white blood cells were reduced. Some patients were also diagnosed with gastrointestinal complications of COVID-19 disease. In patients with sickle cell anemia, a background of chronic inflammation has been identified as a protective factor against COVID-19 disease (13). The mean age of patients was 52.9 years, and 92% of patients were of African descent. Fever and shortness of breath and loss of sense of smell and taste were typical symptoms of patients. They needed supportive care and were also treated with hydroxyl chlorine and steroids. Some patients underwent oxygen therapy and were transferred to the ICU (11). Half of the hospitalized patients required mechanical ventilation, and approximately more than 15% required the ICU. There was a relationship between patients' age and ICU admission. More than half of the hospitalized patients over 45 years of age needed to be hospitalized in the ICU. Older age and vasoconstriction crisis were identified as risk factors for the disease (10). Mutations in the beta chain in hemoglobin may prevent SARS-CoVR binding to iron cells (11).

Since children with sickle cell anemia are more vulnerable to COVID-19 disease, they should wear a mask and maintain social distance (6). Patients with sickle cell anemia often need to receive blood. During the COVID-19 disease pandemic, Blood transfusion centers must follow health protocols. Blood donors should be screened for COVID-19 disease (13). At this time, non-urgent procedures such as bone marrow transplantation should be postponed (17). Complications of sickle cell disease are permanent and temporary defects of the immune system, and the main symptom is fever. Every patient with this symptom should also be screened for COVID-19, regardless of whether they are exposed to the Coronavirus (16).

Health care planning should be considered at the time of COVID19 disease, especially in Africa. Also, special attention should be paid to patients with sickle cell anemia and

their treatment needs (18).

#### 4. Conclusions

COVID 19 is rapidly spreading and is more severe in people with chronic disease and defective immune systems. At the time of producing the appropriate vaccine for COVID 19 disease, patients with sickle cell anemia should be given priority as a high-risk group. Due to the high cost of treating sickle cell disease, international support is necessary, especially during the COVID 19.

#### Footnotes

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#### References

- Hui DS, I. Azhar E, Madani TA, Ntoumi F, Kock R, Dar O, et al. The continuing 2019-nCoV epidemic threat of novel coronaviruses to global health - The latest 2019 novel coronavirus outbreak in Wuhan, China. *Int J Infect Dis.* 2020;**91**:264–6. doi: [10.1016/j.ijid.2020.01.009](https://doi.org/10.1016/j.ijid.2020.01.009). [PubMed: [31953166](https://pubmed.ncbi.nlm.nih.gov/31953166/)]. [PubMed Central: [PMC7128332](https://pubmed.ncbi.nlm.nih.gov/PMC7128332/)].
- She J, Jiang J, Ye L, Hu L, Bai C, Song Y. 2019 novel coronavirus of pneumonia in Wuhan, China: emerging attack and management strategies. *Clin Transl Med.* 2020;**9**(1):19. doi: [10.1186/s40169-020-00271-z](https://doi.org/10.1186/s40169-020-00271-z). [PubMed: [32078069](https://pubmed.ncbi.nlm.nih.gov/32078069/)]. [PubMed Central: [PMC7033263](https://pubmed.ncbi.nlm.nih.gov/PMC7033263/)].
- Wang C, Horby PW, Hayden FG, Gao GF. A novel coronavirus outbreak of global health concern. *Lancet.* 2020;**395**(10223):470–3. doi: [10.1016/s0140-6736\(20\)30185-9](https://doi.org/10.1016/s0140-6736(20)30185-9).
- Wang W, Tang J, Wei F. Updated understanding of the outbreak of 2019 novel coronavirus (2019-nCoV) in Wuhan, China. *J Med Virol.* 2020;**92**(4):441–7. doi: [10.1002/jmv.25689](https://doi.org/10.1002/jmv.25689). [PubMed: [31994742](https://pubmed.ncbi.nlm.nih.gov/31994742/)]. [PubMed Central: [PMC7167192](https://pubmed.ncbi.nlm.nih.gov/PMC7167192/)].
- Zumla A, Niederman MS. Editorial: The explosive epidemic outbreak of novel coronavirus disease 2019 (COVID-19) and the persistent threat of respiratory tract infectious diseases to global health security. *Curr Opin Pulm Med.* 2020;**26**(3):193–6. doi: [10.1097/MCP.0000000000000676](https://doi.org/10.1097/MCP.0000000000000676). [PubMed: [32132379](https://pubmed.ncbi.nlm.nih.gov/32132379/)]. [PubMed Central: [PMC7147276](https://pubmed.ncbi.nlm.nih.gov/PMC7147276/)].
- Vives Corrons JL, De Sanctis V. Rare Anaemias, Sickle-Cell Disease and COVID-19. *Acta Biomed.* 2020;**91**(2):216–7. doi: [10.23750/abm.v91i2.9532](https://doi.org/10.23750/abm.v91i2.9532). [PubMed: [32420949](https://pubmed.ncbi.nlm.nih.gov/32420949/)].
- Muscogiuri G, Pugliese G, Barrea L, Savastano S, Colao A. Commentary: Obesity: The "Achilles heel" for COVID-19? *Metabolism.* 2020;**108**:154251. doi: [10.1016/j.metabol.2020.154251](https://doi.org/10.1016/j.metabol.2020.154251). [PubMed: [32353356](https://pubmed.ncbi.nlm.nih.gov/32353356/)]. [PubMed Central: [PMC7184987](https://pubmed.ncbi.nlm.nih.gov/PMC7184987/)].
- Alberca RW, Oliveira LM, Branco A, Pereira NZ, Sato MN. Obesity as a risk factor for COVID-19: an overview. *Crit Rev Food Sci Nutr.* 2020:1–15. doi: [10.1080/10408398.2020.1775446](https://doi.org/10.1080/10408398.2020.1775446). [PubMed: [32539446](https://pubmed.ncbi.nlm.nih.gov/32539446/)].
- Pettit NN, MacKenzie EL, Ridgway JP, Pursell K, Ash D, Patel B, et al. Obesity is Associated with Increased Risk for Mortality Among Hospitalized Patients with COVID-19. *Obesity (Silver Spring).* 2020;**28**(10):1806–10. doi: [10.1002/oby.22941](https://doi.org/10.1002/oby.22941). [PubMed: [32589784](https://pubmed.ncbi.nlm.nih.gov/32589784/)]. [PubMed Central: [PMC7362135](https://pubmed.ncbi.nlm.nih.gov/PMC7362135/)].
- Arlet J, de Luna G, Khimoud D, Odièvre M, de Montalembert M, Joseph L, et al. Prognosis of patients with sickle cell disease and COVID-19: a French experience. *Lancet Haematol.* 2020;**7**(9):e632–4. doi: [10.1016/s2352-3026\(20\)30204-0](https://doi.org/10.1016/s2352-3026(20)30204-0).

11. Balanchivadze N, Kudirka AA, Askar S, Almadhoun K, Kuriakose P, Fadel R, et al. Impact of COVID-19 Infection on 24 Patients with Sickle Cell Disease. One Center Urban Experience, Detroit, MI, USA. *Hemoglobin*. 2020;1-6. doi: [10.1080/03630269.2020.1797775](https://doi.org/10.1080/03630269.2020.1797775). [PubMed: [32722950](https://pubmed.ncbi.nlm.nih.gov/32722950/)].
12. Heilbronner C, Berteloot L, Tremolieres P, Dupic L, de Saint Blanquat L, Lesage F, et al. Patients with sickle cell disease and suspected COVID-19 in a paediatric intensive care unit. *Br J Haematol*. 2020;190(1):e21-4. doi: [10.1111/bjh.16802](https://doi.org/10.1111/bjh.16802). [PubMed: [32420608](https://pubmed.ncbi.nlm.nih.gov/32420608/)]. [PubMed Central: [PMC7276717](https://pubmed.ncbi.nlm.nih.gov/PMC7276717/)].
13. Hussain FA, Njoku FU, Saraf SL, Molokie RE, Gordeuk VR, Han J. COVID-19 infection in patients with sickle cell disease. *Br J Haematol*. 2020;189(5):851-2. doi: [10.1111/bjh.16734](https://doi.org/10.1111/bjh.16734). [PubMed: [32314798](https://pubmed.ncbi.nlm.nih.gov/32314798/)]. [PubMed Central: [PMC7264585](https://pubmed.ncbi.nlm.nih.gov/PMC7264585/)].
14. Justino CC, Campanharo FF, Augusto MN, Morais SC, Figueiredo MS. COVID-19 as a trigger of acute chest syndrome in a pregnant woman with sickle cell anemia. *Hematol Transfus Cell Ther*. 2020;42(3):212-4. doi: [10.1016/j.htct.2020.06.003](https://doi.org/10.1016/j.htct.2020.06.003). [PubMed: [32646838](https://pubmed.ncbi.nlm.nih.gov/32646838/)]. [PubMed Central: [PMC7301062](https://pubmed.ncbi.nlm.nih.gov/PMC7301062/)].
15. Taher AT, Bou-Fakhredin R, Kreidieh F, Motta I, De Franceschi L, Cappellini MD. Care of patients with hemoglobin disorders during the COVID-19 pandemic: An overview of recommendations. *Am J Hematol*. 2020;95(8):E208-10. doi: [10.1002/ajh.25857](https://doi.org/10.1002/ajh.25857). [PubMed: [32394480](https://pubmed.ncbi.nlm.nih.gov/32394480/)]. [PubMed Central: [PMC7272998](https://pubmed.ncbi.nlm.nih.gov/PMC7272998/)].
16. John NA, John JE. Implications of COVID-19 infections in sickle cell disease. *Pan Afr Med J*. 2020;36:81. doi: [10.11604/pamj.2020.36.81.23776](https://doi.org/10.11604/pamj.2020.36.81.23776). [PubMed: [32774640](https://pubmed.ncbi.nlm.nih.gov/32774640/)]. [PubMed Central: [PMC7386278](https://pubmed.ncbi.nlm.nih.gov/PMC7386278/)].
17. Roy NBA, Telfer P, Eleftheriou P, de la Fuente J, Drasar E, Shah F, et al. Protecting vulnerable patients with inherited anaemias from unnecessary death during the COVID-19 pandemic. *Br J Haematol*. 2020;189(4):635-9. doi: [10.1111/bjh.16687](https://doi.org/10.1111/bjh.16687). [PubMed: [32330288](https://pubmed.ncbi.nlm.nih.gov/32330288/)]. [PubMed Central: [PMC7264776](https://pubmed.ncbi.nlm.nih.gov/PMC7264776/)].
18. Dexter D, Simons D, Kiyaga C, Kapata N, Ntoumi F, Kock R, et al. Mitigating the effect of the COVID-19 pandemic on sickle cell disease services in African countries. *Lancet Haematol*. 2020;7(6):e430-2. doi: [10.1016/s2352-3026\(20\)30122-8](https://doi.org/10.1016/s2352-3026(20)30122-8).