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Sickle cell anemia and COVID-19: Use of voxelotor to avoid transfusion

To the Editor

The American Society of Hematology (ASH) has provided guidance on the use of red blood cell transfusions in patients with sickle cell disease (SCD) in the setting of COVID-19, as blood donations have dropped off significantly in some regions of the United States.[1] ASH reports that these blood shortages may require modification in the thresholds for transfusion for common manifestations of SCD such as anemia, vaso-occlusive crisis, and priapism, although they recommend the continuation of transfusion guidelines for acute complications such as stroke or acute chest syndrome. During this period, hospitals have been required to carefully manage their blood supplies as the shortages persist.[2] The shortages may require loosening exchange transfusion endpoints (ie, targeting 40% hemoglobin S instead of 30%) or switching to simple transfusion for patients with SCD. Additionally, the AABB, the American Red Cross, and America's Blood Centers recently released a statement confirming that blood inventories are still decreasing as the pandemic continues.[3] Here we report on a patient with SCD that experienced a significant drop in hemoglobin during hospitalization for COVID-19 infection who was successfully treated with voxelotor instead of additional transfusions.

A 39-year-old female with HbSS SCD was admitted to the hospital with diffuse skeletal pain that was not relieved by ibuprofen and later oxycodone at home. She had been compliant with COVID-19 social distancing restrictions, isolating with her husband and son at home, and had no recent illness or exposure to sick contacts. In the emergency department, she was noted to be afebrile with stable vital signs and with oxygen saturation of 97% on room air. Laboratory studies were notable for WBC 12.6 K/uL, hemoglobin (Hb) of 7.9 g/dL, reticulocyte count of 12.9%, total bilirubin 7.5 mg/dL, and d-dimer of 3.55 ug/mL. The patient had type O/Rh + blood and no history of alloantibodies. Chest X-ray revealed stable cardiomegaly and coarse pulmonary markings. Diagnosed with acute sickle crisis, she was treated with morphine by patient-controlled analgesia (PCA) and IV saline. Shortly after admission, she spiked a fever to 101.7°F and became hypotensive. Nasopharyngeal swab for SARS-CoV-2 (COVID-19) was positive by PCR (Abbott). She became short of

TABLE 1 Laboratory parameters [Color table can be viewed at wileyonlinelibrary.com]

Date/2020	3/16[a]	4/28[b]	4/29	4/30	5/1	5/2	5/3	5/4	5/5	5/6	5/7[c]	5/12[d]	5/19[d]
Voxelotor treatment													
Hb, g/dL	7.6	7.9	6.7	6.9	6.7	6.9	7.0	6.5	6.4	8.0	8.1	10.3	10.4
Retic Ct (%)		12.9								20.2	17.3	6.5	
Ind Bili, mg/dL		4.1				3.7	3.3	2.0	3.4	1.4		0.8	

Abbreviations: Hb, hemoglobin (11.4-14.8 g/dL); Ind Bili, indirect bilirubin (0.2-1.0 mg/dL); Retic Ct, reticulocyte count (0.8-2.3%).
[a]Clinic visit 5 weeks prior to hospitalization. Hb in her typical range of 7.5-8.5 g/dL.
[b]Admitted with sickle crisis and COVID pneumonia.
[c]Discharged home, clinically stable, improved respiratory status.
[d]Clinic visits 5 and 12 days after discharge.

breath and was treated with oxygen to maintain oxygen saturation >90%. Her hypoxia was attributed to COVID-19. At the time of her admission, there were no COVID-19-specific trials active at our institution, and she received no specific COVID intervention.

Hb had fallen to 6.7 g/dL and she was transfused with 2 units of leukoreduced, fully cross-matched red blood cells (RBCs), but without a resulting increase in Hb level. Post hydration volume redistribution and hyperhemolysis were considered as potential causes of this lack of response. No alloantibodies were detected at the time of transfusion.

Treatment with erythrocytapheresis which requires nurse and technician presence for 4 to 5 hours was contemplated for this patient. However, the patient did not meet the criteria for acute chest syndrome; she had improved hemodynamically, and in an effort to avoid additional transfusions and the associated health care provider exposure risks during a period of limited personal protective equipment, we elected to administer voxelotor 1500 mg orally daily. Voxelotor is a HbS polymerization inhibitor that increases hemoglobin in individuals with SCD, thus improving oxygen carrying capacity.[4,5] Within 2 days of starting voxelotor, her Hb had risen to 8.0 g/dL (Table 1). She remained clinically stable and was discharged home off supplemental oxygen (room air O2 sat 98%). By day 10, her Hb was 10.3 g/dL. To our knowledge, this is the first report of voxelotor being used acutely in the setting of COVID-19 pneumonia in an individual with SCD and respiratory distress in lieu of transfusion. In this case, the patient's Hb and overall status improved quickly upon treatment, thereby avoiding exchange transfusion, decreasing hospital staff exposure to coronavirus and sparing RBC units; important during this pandemic era of limited blood supply.

CONFLICT OF INTEREST

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Association of ABO blood group and secretor phenotype with severe COVID-19

To the Editor

We read with interest the letter by Dzik and colleagues,[1] who compared the ABO blood group distribution between 957 COVID-19 patients admitted in the Boston area during the 2020 pandemic for whom ABO typing was available and 5840 historical controls from the same period of 2019 and found no significant difference. The investigation was prompted by evidence from previous studies conducted in Asia and Mediterranean Europe highlighting a higher risk of severe SARS-CoV-2