

Sickle Cell Anemia and COVID-19: Use of Voxelotor to Avoid Transfusion

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- She had type O/Rh+ blood and no history of alloantibodies. Laboratory results were notable for WBC count (12.6 K/ μ L), Hb (7.9 g/dL), reticulocyte count (12.9%), total bilirubin (7.5 mg/dL), and D-dimer (3.55 μ g/mL) (**Table 1**).

Table 1: Laboratory Parameters

	Prior Clinic Visit	Admission	Hospitalization									Discharged	Follow-up Visits		
			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	
Date (2020)	3/16 ^a	4/28 ^b											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		
Reticulocyte count, %		12.9								20.2	17.3	6.5			
Indirect bilirubin, mg/dL		4.1				3.7	3.3	2.0	3.4	1.4	0.8				

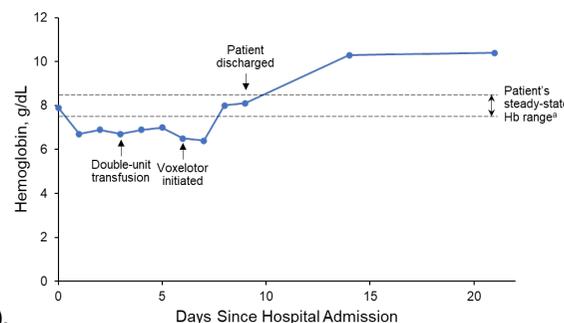
Abbreviations: Hb, hemoglobin

Normal ranges: Hb, 11.4-14.8 g/dL; reticulocyte count, 0.8%-2.3%; indirect bilirubin, 0.2-1.0 mg/dL.

^aClinic visit 5 weeks before hospitalization. Hb is in the patient's typical range of 7.5 to 8.5 g/dL. ^bPatient admitted with sickle crisis and COVID-19 pneumonia. ^cPatient discharged, clinically stable with improved respiratory status; ^dClinic visits 5 and 12 days after discharge.

- Chest X-ray revealed stable cardiomegaly and coarse pulmonary markings. The patient was diagnosed with acute sickle cell crisis and treated with morphine via patient-controlled analgesia and intravenous saline.
- Shortly after being admitted, her temperature spiked to 101.7 °F, and she became hypotensive.
- A nasopharyngeal swab sample was confirmed positive for SARS-CoV-2 (COVID-19) via PCR (Abbott).
 - The patient became short of breath and was given oxygen to maintain oxygen saturation >90%.
- After her Hb level fell to 6.7 g/dL, she was transfused with 2 units of fully cross-matched, leukoreduced RBCs; no increase in Hb was observed, and no alloantibodies were detected at the time of transfusion.

Figure 1. Change in the Patient Hemoglobin Levels Since Admission to Hospital



Abbreviations: Hb, hemoglobin

^aThe patient's Hb typical range is 7.5 to 8.5 g/dL.

BACKGROUND

- The COVID-19 pandemic has severely disrupted blood donations and created a critical shortage in the US blood supply.¹
- Many patients with sickle cell disease (SCD) rely on red blood cell (RBC) transfusions as prophylactic treatment or for the management of SCD-related complications.²
- As the pandemic persists, hospitals must cautiously manage their blood supplies.¹
 - The American Society of Hematology has released guidance for the use of RBC transfusions in patients with SCD in the context of COVID-19.³
- Voxelotor is a sickle hemoglobin–polymerization inhibitor approved in the United States for treatment for SCD in patients aged ≥ 12 years that increases hemoglobin (Hb) in individuals with SCD, improving oxygen carrying capacity.^{4,5}
- Here we describe the case of a patient with SCD who showed a significant drop in Hb while being hospitalized for COVID-19 infection. With use of voxelotor, rather than additional transfusions, the patient was treated successfully.

CASE DESCRIPTION

- A 39-year-old female with HbSS SCD was admitted to the hospital with diffuse skeletal pain that was not alleviated with ibuprofen or oxycodone taken at home.
- While in the emergency department, she was afebrile with stable vital signs and oxygen saturation of 97% on room air.

- Treatment with erythrocytapheresis was contemplated, which would have required attendance of a nurse and technician for 4 to 5 hours. However, the patient did not meet the criteria for acute chest syndrome, and she had improved hemodynamically.
- In an effort to avoid additional transfusions and any associated exposure risk for hospital staff, with personal protective equipment being limited, the patient was administered voxelotor 1500 mg orally once daily.
 - Within 2 days of initiating voxelotor treatment, the patient's Hb increased to 8.0 g/dL (**Figure 1**).
- She remained clinically stable and was discharged home, without supplemental oxygen (room air oxygen saturation was 98%).
 - After 10 days of treatment, her Hb was 10.3 g/dL.

CONCLUSIONS

- The patient's Hb and overall status were improved quickly with voxelotor treatment, thereby avoiding exchange transfusion, sparing RBC units, and decreasing exposure of healthcare providers to COVID-19, all of which are important considerations during this era of pandemic and limited blood supply.

References: 1. America's Blood Centers. America's Blood Centers, AABB and American Red Cross Release Joint Statement on the Blood Supply. Accessed September 30, 2020. <https://americasblood.org/news/joint-statement-on-the-blood-supply/>. 2. National Heart, Lung, and Blood Institute. Evidence-based management of sickle cell disease: expert panel report, 2014. Accessed September 30, 2020. <https://www.nhlbi.nih.gov/health-topics/evidence-based-management-sickle-cell-disease>. 3. American Society of Hematology. COVID-19 and Sickle Cell Disease: Frequently Asked Questions. Accessed September 30, 2020. <https://www.hematology.org/covid-19/covid-19-and-sickle-cell-disease>. 4. Oxbrta [package insert]. South San Francisco, CA: Global Blood Therapeutics; 2019. 5. Vichinsky E, Hoppe CC, Ataga KI, et al. A phase 3 randomized trial of voxelotor in sickle cell disease. *N Engl J Med*. 2019;381(6):509-519.

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