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

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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 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 afibrile with stable vital signs and oxygen saturation >90%.
- She had type O/Rh+ blood and no history of alloantibodies. Laboratory results were notable for WBC count (12.6 K/µL), Hb (7.9 g/dL), reticulocyte count (12.9%), total bilirubin (7.5 mg/dL), and D-dimer (3.55 µg/mL) (Table 1).
- She had type O/Rh+ blood and no history of alloantibodies. Laboratory results were notable for WBC count (12.6 K/µL), Hb (7.9 g/dL), reticulocyte count (12.9%), total bilirubin (7.5 mg/dL), and D-dimer (3.55 µg/mL) (Table 1).
- Chest X-ray revealed stable cardiomegaly and coarse pulmonary markings. The patient was diagnosed with acute sickle 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.
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- 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.

Disclosures
- We thank the patient and hospital staff who contributed to this report.
- William B. Ershler: Global Blood Therapeutics, Novartis, consultant.
  - Margaret E. Holbrook: Nothing to disclose.

References

Table 1: Laboratory Parameters

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<th>Prior Clinic Visit</th>
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<th>Follow-up Visits</th>
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<td>Voxelotor treatment</td>
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<td>Indirect bilirubin</td>
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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.

Acknowledgments
- We thank the patient and hospital staff who contributed to this report.

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