

Sickle Cell Disease Patients Presenting to the Emergency Department During the COVID-19 Pandemic Considerations and a Checklist

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Introduction

Symptoms concerning for COVID-19, including fever, dyspnea, cough and pain, overlap with those resulting from serious SCD complications, including acute chest syndrome, pulmonary embolus (PE) and cardiac failure/pulmonary hypertension. It is important to evaluate and treat these patients not only for a presumed COVID-19 etiology, but also for concurrent or alternative SCD complications as explanations for symptoms. In addition, the most common presenting symptom for SCD with COVID-19 disease is pain. Preliminary data suggests that SCD patients may have higher mortality from COVID-19 due to underlying predisposing conditions related to SCD (cardiac diastolic dysfunction, asthma, pulmonary hypertension, cerebrovascular disease, renal dysfunction, thrombotic risk) and/or severe SCD complications such as acute chest syndrome or PE secondary to COVID-19 infection. Even patients with “milder” phenotypes such as Hb SC disease can develop respiratory failure and other severe complications concurrent with COVID-19 infection.

The following checklist is intended to assist in the evaluation of SCD patients in the ED presenting with symptoms concerning for COVID-19 and/or complications of SCD, with SCD-specific considerations for evaluation and treatment.

Clinical and laboratory evaluation:

- SARS-CoV-2 viral testing (+/- other respiratory virus/influenza testing depending on institutional practice and seasonal considerations)
- CXR for any respiratory symptoms even in absence of hypoxemia or lung findings on exam
 - Consider V/Q scan or CT-PA scan if PE is suspected
- Electrocardiogram, troponin in those with chest pain, dyspnea or cardiac history
- CBC with differential, screening for anemia and thrombocytopenia
- Reticulocyte count (given concern for B19 aplastic crisis if low, hyper-hemolysis if very high)
- D-dimers-marked elevation suggests serious COVID or VTE or both and may suggest need for CT-PA scan, venous Dopplers and admission
- Electrolytes, BUN/creatinine, bilirubin, liver enzymes, LDH given possible underlying kidney and liver dysfunction linked to SCD and COVID
- Blood cultures if febrile, hypotensive or toxic-appearing: SCD patients are especially prone to infection with encapsulated organisms. Sputum cultures often uninformative and risky to obtain during pandemic
- Procalcitonin, urinary Strep Pneumoniae and Legionella testing if available
- Type and screen all patients on presentation to prepare for possible simple or exchange transfusion

- Must include extended RBC antigen typing for C, E and Kell given high rate of alloimmunization in SCD
- Consult hematology and blood bank if transfusion needed

Interventions:

- Consultation with hematologist in ED, if possible
- Consultation with obstetrics if SCD patient is pregnant
- Supplementary oxygen only if hypoxic (<94% or >4% below baseline if have chronic hypoxia) to raise pO₂ >94%
- Judicious fluid replacement with D5W if signs/symptoms of fluid deficit,
 - No more than 125ml/hour, decrease to 50ml/hour after 1 liter if tolerating oral fluids
 - Avoid fluid bolus as this may exacerbate pulmonary edema in SCD patients
- Empiric antibiotics until bacterial infection ruled out if persistent high fever/unstable vital signs
 - For suspected pneumonia: macrolide and a third generation cephalosporin
 - Vancomycin if concern for line or skin infection (SCD patients can have an indwelling port or line, and/or leg ulcers)
- Transfuse to decrease HbS and treat possible acute chest syndrome, particularly if Hb > 2gr/dL below baseline or serious respiratory compromise
 - Begin with simple transfusion if Hb < 10gr/dL
 - If Hb > 10gr/dL, given concern for hyperviscosity, arrange exchange transfusion
 - Consult hematology immediately if considering transfusion
- IV pain medication as needed, following pre-established individualized plan or institutional protocols

Admit any SCD patient with evidence for ACS, pneumonia, worsening or severe hypoxemia or non-controlled pain for observation, aggressive IV pain management, antibiotics until cultures negative, and oxygen as needed:

- In resource-limited care settings without access to pulse oximetry, a RR>24 should be cause for concern and potential admission

If patient is discharged from ED:

- Arrange telemedicine or in-person visit with hematologist or primary SCD provider within 24 hours
- Stress low threshold for return to ER and admission if symptoms, especially SOB, worsen
- Provide pulse oximeter if feasible

For additional information, see:

- ASH COVID and SCD FAQs
<https://www.hematology.org/covid-19/covid-19-and-sickle-cell-disease>
- An outline to decrease burden and minimize morbidity from COVID-19 in SCD
<https://www.sicklecelldisease.org/2020/03/18/sickle-cell-disease-and-covid-19-provider-directory/>
- Registry for reporting of patients with COVID-19 and SCD
<https://covidsicklecell.org/updates-data/>

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