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
- An outline to decrease burden and minimize morbidity from COVID-19 in SCD
- Registry for reporting of patients with COVID-19 and SCD
  https://covidssicklecell.org/updates-data/

Please note:

While ASH aims to provide a forum for the exchange of information that is helpful, accurate and up-to-date, the relevant health information and data concerning the disease, as well as the various governmental actions and responses to the disease, are changing constantly, and ASH makes no representation, warranty, or guaranty whatsoever as to any information that may be exchanged through this webpage.

It remains the responsibility of each medical professional to decide on the appropriate medical advice, diagnosis, and treatment for patients. In rendering such advice, please bear in mind that although ASH attempts to include on this page information from reliable sources, it has not conducted a substantive review of each item posted below, nor does it have access to sufficient information to police potential conflicts of interest. Particularly given the exigencies presented by the current COVID-19 pandemic, we urge you to assist us identifying information that has been particularly helpful to you so that we might more prominently feature it, as well as any information that is inaccurate, outdated or inappropriate so that we can remove it.