7. Dong Y, Mo X, Hu Y, Qi X, Jiang F, Jiang Z, et al. Epidemiological characteristics of 2143 pediatric patients with 2019 coronavirus disease in China. *Pediatrics*. 2020. https://doi.org/10.1542/peds.2020-0702

8. Kam KQ, Yung CF, Lin RTP, Mak TM, Maiwald M, Li J, et al. A well infant with Coronavirus disease 2019 (COVID-19) with high viral load. *Clin Infect Dis*. 2020. https://doi.org/10.1093/cid/ciaa201

COVID-19 infection in patients with sickle cell disease

Severe acute respiratory syndrome coronavirus 2, also known as COVID-19, has spread to 184 countries, with almost 1·5 million cases (as of mid-April 2020) since first reported.[1] The clinical features of this disease are not completely understood; however, severe illness is thought to occur predominantly in adults with advanced age and those with underlying comorbidities.[2] Sickle cell disease (SCD), an immunocompromised condition, puts patients at higher risk for respiratory infections and subsequent pulmonary complications such as acute chest syndrome (ACS).[3] Here, we present a case series of four SCD patients who were found to be positive for COVID-19 and describe our approach to management.

Case 1

A 32-year-old male, haemoglobin (Hb) SS, history of recurrent vaso-occlussive crises (VOC), ACS and chronic lower extremity ulcers presented to the emergency department (ED) with a typical VOC. He was afebrile and his pulse oximetry was 97% in room air. After admission, he was treated with intravenous (IV) morphine and fluids. He had a fever of 38·5°C on the second day of hospitalisation. A nasopharyngeal swab ordered because of a dry cough and a sore throat was COVID-19 positive, and a chest x-ray (CXR) that day showed plate-like atelectasis above the left lower lobe, suggesting developing pneumonia. He was treated with ceftriaxone (later changed to piperacillin-tazobactam) and azithromycin for seven days, along with hydroxychloroquine (200 mg twice daily) throughout the hospital stay. Starting the second day of this hospitalisation, supplementary oxygen via nasal cannula was initiated at two litres per minute after the oxygen saturation declined to 88% in room air, increased to four litres per minute later on and subsequently was intubated due to increasing oxygen requirements in the intensive care unit (ICU). After receiving a simple transfusion (1 unit) followed by an exchange blood transfusion in the ICU, his condition started to improve. He was extubated after 4 days and discharged home after 13 days of hospitalisation (Table I).

Case 2

A 37-year-old female, HbSβ^+, with a history of ACS, frequent VOC and venous thromboembolism presented to the ED with typical VOC pain. She was afebrile in the ED with a negative CXR after complaining of subjective fevers at home. After admission, she was treated with IV morphine and fluids, but a nasopharyngeal swab ordered due to upper respiratory symptoms (nasal congestion) was positive for COVID-19. During the hospitalisation, her pain gradually improved. She remained afebrile and did not require any supplementary oxygen. Her laboratory values were stable, with the exception of her white blood cell (WBC) count declining to a nadir of $2{\cdot}3 \times 10^3/\mu l$. She has discharged home after 8 days.

Case 3

A 22-year-old female, HbSS, with a history of ACS, frequent VOC and asthma presented to the ED with severe pain and nausea, vomiting and diarrhoea for one day. She was afebrile in ED, but had one episode of fever to 39·25°C after admission. The COVID-19 test, ordered due to the gastrointestinal symptoms, was positive. She was treated with IV morphine and started on ceftriaxone for suspected ACS due to the fever episode, although she had stable oxygen saturations (>95%). She defervesced on the second day of hospitalisation, her pain gradually improved and she was discharged after 2 days of stay.

Case 4

A 41-year-old male, HbSC, with history of bilateral hip avascular necrosis (AVN) and a pulmonary embolism (PE) presented to the ED with worsening hip pain. He developed a cough and dyspnea one week earlier and was diagnosed with COVID-19 infection at another institution, but left against medical advice. After admission he was treated with IV morphine and fluids for a VOC. Throughout this hospitalisation, he was afebrile and did not develop any respiratory symptoms. His pain continued to improve and he was discharged after 4 days.

It is reported that the COVID-19 infection disproportionately affects more African Americans than other ancestries,[4] which may impose a higher risk in patients with SCD, especially considering the compromised immune system of this patient population. We tested 14 SCD patients for COVID-19 and three tested positive at our institution. Despite different SCD genotypes, the four patients all have history of

Table I. Clinical presentation of patient cases.

Patient case	Age	Gender	SCD genotype	PMH	Initial temperature	Highest temperature	Initial O2sat	Supplementary O2 requirement	Initial WBC (10^3/ul)	Zenith WBC (10^3/ul)	Initial Hgb (g/l)	Nadir Hgb (g/l)	CXR	LOS (days)	Management
Patient #1	32	M	HbSS	ACS	36·8	39·3	97%	Yes	14·3	22·7	8·2	7·3	Positive	13	IV pain meds, antibiotics, hydroxychloroquine, supplemental oxygen, intubation, exchange transfusion
Patient #2	37	F	HbSβ+	ACS	37·3	37·5	100%	No	5·3	5·3	12·2	10·1	Negative	8	IV pain meds
Patient #3	22	F	HbSS	ACS, asthma	36·9	39·2	100%	No	10·9	16·0	8·1	7·3	Negative	2	IV pain meds, antibiotics
Patient #4	41	M	HbSC	PE	37·1	38·0	98%	No	7·6	8·1	12·1	11·4	Negative	4	IV pain meds

respiratory complications, such as ACS, asthma or PE, which may be potential risk factors for progressive COVID-19 pulmonary disease in patients with SCD. All four patients initially presented to the ED for typical VOC, and the clinical course of their COVID-19 infection was rather mild considering these patients were immunocompromised (except for the patient in Case 1, where the ACS episode may or may not have been related to the COVID-19 infection). We wonder if the background chronic inflammatory, haemolytic and anaemic state in SCD might have a favourable influence in protecting this patient population from fatal COVID-19 infection. Severe respiratory symptoms due to COVID-19 would qualify for the definition of ACS in SCD patients, and we believe that red blood cell exchange transfusion should be urgently performed. Further studies with more patients will be needed for a better understanding of COVID-19 in this patient population.

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References

1. https://coronavirus.jhu.edu/map.html. Last accessed on April 9, 2020.
2. Preliminary estimates of the prevalence of selected underlying health conditions among patients with coronavirus disease 2019 — United States, February 12–March 28, 2020. *MMWR Morb Mortal Wkly Rep*. 2020;**69**:382–6.
3. Steinberg MH. Management of sickle cell disease. *N Engl J Med*. 1999;**340** (13):1021–30.
4. Garg SKL, Whitaker M, et al. Hospitalization rates and characteristics of patients hospitalized with laboratory-confirmed coronavirus disease 2019 — COVID-NET, 14 States, March 1–30, 2020. MMWR *Morb Mortal Wkly Rep*. ePub: 8 April 2020.