# Evidence Search Service Results of your search request

## COVID risk for people with Sickle cell disease

**ID of request:** 28702  
**Date of request:** 7th April, 2021  
**Date of completion:** 9th April, 2021

If you would like to request any articles or any further help, please contact:  Igor Brbre at [igor.brbre@nhs.net](mailto:igor.brbre@nhs.net)

Please acknowledge this work in any resulting paper or presentation as: Evidence search: COVID risk for people with Sickle cell disease. Igor Brbre. ( 9th April, 2021). BRIGHTON, UK: Brighton and Sussex Library and Knowledge Service.

**Sources searched**  
Cochrane Library (0)  
EMBASE (20)  
Europe PubMed Central (5)  
Google Scholar (0)  
MEDLINE (37)

**Date range used** (5 years, 10 years): no restrictions   
**Limits used** (gender, article/study type, etc.): none   
**Search terms and notes** (full search strategy for database searches below):

Relevant natural language and controlled vocabulary terms were selected and combined. Thesaurus terms were adapted for different databases. Medline and Embase searched on Ovid. Results were reviewed for relevance and de-duplicated in EndNote. Full search strategy below.

Preprints search in Europe PMC:

(("COVID-19" or COVID19 or 2019nCoV or "Corona Virus" or Coronavirus or "CoV 2" or CoV2 or COVID or nCoV or SARS2 or SARSCoV or "SARS-CoV") AND ("Sickle Cell" AND (anemia or anaemia or disorder\* or disease or illness)) AND (risk or probability or likelihood or prevalence or incidence)) AND (SRC:PPR)

For more information about the resources please go to: <https://www.bsuh.nhs.uk/library/>.

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## A. Systematic Reviews

#### Cochrane Database of Systematic Reviews

**Prophylactic antibiotics for preventing pneumococcal infection in children with sickle cell disease** (2021)

Rankine-Mullings Angela E., Owusu-Ofori Shirley

[Available online at this link](https://www.knowledgeshare.nhs.uk/index.php?PageID=link_resolver&link=19b8013e857cb0dc84905ef4c7c8891a)

BACKGROUND: Sickle cell disease (SCD) is a group of inherited disorders that result in haemoglobin abnormalities and other complications. Injury to the spleen, among other factors, contribute to persons with SCD being particularly susceptible to infection. Infants and very young children are especially vulnerable. The 'Co-operative Study of Sickle Cell Disease' observed an incidence rate for pneumococcal septicaemia of 10 per 100 person-years in children under the age of three years. Vaccines, including customary pneumococcal vaccines, may be of limited use in this age group. Therefore, prophylactic penicillin regimens may be advisable for this population. This is an update of a Cochrane Review which was first published in 2002, and previously updated, most recently in 2017. OBJECTIVES: To compare the effects of antibiotic prophylaxis against pneumococcus in children with SCD receiving antibiotic prophylaxis compared to those without in relation to: 1. incidence of Streptococcus pneumoniae infection; 2. mortality (as reported in the included studies); 3. drug-related adverse events (as reported in the included studies) to the individual and the community; 4. the impact of discontinuing at various ages on incidence of infection and mortality., SEARCH METHODS: We searched the Cochrane Cystic Fibrosis and Genetic Disorders Group Haemoglobinopathies Trials Register, which is comprised of references identified from comprehensive electronic database searches and also two clinical trials registries: ClinicalTrials.gov and the WHO International Registry Platform (not in 2020 given access issues relating to Covid-19 pandemic). Additionally, we carried out hand searching of relevant journals and abstract books of conference proceedings. Date of the most recent search: 25 January 2021., SELECTION CRITERIA: All randomised or quasi-randomised controlled trials comparing prophylactic antibiotics to prevent pneumococcal infection in children with SCD with placebo, no treatment or a comparator drug., DATA COLLECTION AND ANALYSIS: The standard methodological procedures expected by Cochrane were used. Both authors independently extracted data and assessed trial quality. The authors used the GRADE criteria to assess the certainty of the evidence., MAIN RESULTS: Six trials were identified by the searches, of which three trials were eligible for inclusion. A total of 880 children, who were between three months to five years of age at randomization were included. The included studies were conducted in centres in the USA and in Kingston, Jamaica. In trials that investigated initiation of penicillin on risk of pneumococcal infection, the odds ratio was 0.37 (95% confidence interval 0.16 to 0.86) (two trials, 457 children) (low-certainty evidence), while for withdrawal the odds ratio was 0.49 (95% confidence interval 0.09 to 2.71) (one trial, 400 children) (low-certainty evidence). Adverse drug effects were rare and minor. Rates of pneumococcal infection were found to be relatively low in children over the age of five years. Overall, the certainty of the evidence for all outcomes was judged to be low. The results from the risk of bias assessment undertaken identified two domains in which the risk of bias was considered to be high, these were incomplete outcome data (attrition bias) (two trials) and allocation concealment (selection bias) (one trial). Domains considered to have a low risk of bias for all three trials were selective reporting (reporting bias) and blinding (performance and detection bias)., AUTHORS' CONCLUSIONS: The evidence examined was determined to be of low certainty and suggests that prophylactic penicillin significantly reduces risk of pneumococcal infection in children with homozygous SCD, and is associated with minimal adverse reactions. Further research may help to determine the ideal age to safely withdraw penicillin. Copyright © 2021 The Cochrane Collaboration. Published by John Wiley & Sons, Ltd.

## B. Original Research

1. **Clinical predictors of poor outcomes in patients with sickle cell disease and COVID-19 infection**  
   Minniti Caterina P. Blood advances 2021;5:207-215.

We aimed to identify predictors of outcomes and survival in patients living in 4 major metropolitan areas who had sickle cell disease (SCD) and COVID-19 to inform best approaches to prevention and care. Data were collected at baseline and during the clinical course in SCD patients diagnosed with COVID-19 in four COVID-19 epicenters. Patients were followed up posthospital discharge for up to 3 months. Of sixty-six SCD patients with COVID-19, fifty patients (75%) required hospitalization, and seven died (10.6%). Patients with preexisting kidney disease (chronic kidney disease) were more likely to be hospitalized. The most common presenting symptom was vaso-occlusive pain. Acute chest syndrome occurred in 30 (60%) of the 50 hospitalized patients and in all who died. Older age and histories of pulmonary hypertension, congestive heart failure, chronic kidney disease, and stroke were more prevalent in patients who died, as were higher creatinine, lactate dehydrogenase, and D-dimer levels. Anticoagulation use while inpatient was twice less common in patients who died. All deaths occurred in individuals not taking hydroxyurea or any other SCD-modifying therapy. Patients with SCD and COVID-19 exhibited a broad range of disease severity. We cannot definitively state that the overall mortality is higher in patients with SCD, although our case fatality rate was ~10% compared with ~3% in the general population, despite a median age of 34 years. Individuals with SCD aged >50 years, with preexisting cardiopulmonary, renal disease, and/or stroke not receiving hydroxyurea, who present with high serum creatinine, lactate dehydrogenase, and D-dimer levels, are at higher risk of death, irrespective of genotype or sex. Copyright © 2021 by The American Society of Hematology.

[Available online at this link](https://www.knowledgeshare.nhs.uk/index.php?PageID=link_resolver&link=eed0cd13325b2f517888bae68d243e79)

1. **Coronavirus Disease 2019 (COVID-19) Infection in Children With Sickle Cell Disease: Case Series From Oman**  
   Al Sabahi Amal Journal of pediatric hematology/oncology 2021;:No page numbers.

BACKGROUND: In March 2020, WHO announced Coronavirus Disease 2019 (COVID-19) outbreak a global pandemic. During this pandemic, patients with sickle cell disease (SCD) have been placed in the "high-risk" category of the population. Although there are numerous publications describing COVID-19 in adult patients, pediatric data are still limited., OBSERVATION: Herein, we report case series of 5 sickle cell disease Omani children who got infected with COVID-19; illustrating their different ways of presentation, management and highlighting the outcomes., CONCLUSION: Although SCD patients are considered as a high-risk group, all of the observed patients, and whose cases are reported here, have recovered. A large scale of SCD cases should be studied to reach more conclusive results. Copyright © 2021 Wolters Kluwer Health, Inc. All rights reserved.

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1. **COVID-19 and Vulnerable Populations in Sub-Saharan Africa**  
   George J. A. Advances in experimental medicine and biology 2021;1321:147-162.

The novel corona virus 2019 (COVID-19) outbreak which started in Hubei province in China has now spread to every corner of the earth. While the pandemic started later in Africa, it is now found in all African countries to varying degrees. It is thought that the prevalence and severity of disease is influenced by a number of non-communicable diseases (NCDs) which are all becoming increasingly prevalent in sub-Saharan Africa (SSA). In addition, SSA bears the major burden of human immunodeficiency virus (HIV) and tuberculosis (TB) infections. While data from Europe and the United States show that children are spared severe disease, it is uncertain if the same holds true in SSA where children suffer from sickle cell disease and malnutrition in addition to other infectious diseases. There is limited data from Africa on the effects of these conditions on COVID-19. In this review, we discuss the epidemiology of some of these conditions in Africa and the possible pathogenesis for the interactions of these with COVID-19.

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1. **COVID-19 in individuals with sickle cell disease/trait compared with other Black individuals**  
   Singh Ashima Blood advances 2021;5:1915-1921.

In the United States, COVID-19 has disproportionately affected Black persons. Sickle cell disease (SCD) and sickle cell trait (SCT) are genetic conditions that occur predominantly among Black individuals. It is unknown if individuals with SCD/SCT are at higher risk of severe COVID-19 illness compared with Black individuals who do not have SCD/SCT. The objective of our study was to compare COVID-19 outcomes, including the disease manifestations, hospitalization, and death, among individuals with SCD/SCT vs Black individuals who do not have SCD/SCT. We leveraged electronic health record data from a multisite research network to identify Black patients with COVID-19 who have SCD/SCT and those who do not have SCD/SCT. During the study period of 20 January 2020 to 20 September 2020, there were 312 patients with COVID-19 and SCD and 449 patients with COVID-19 and SCT. There were 45 517 Black persons who were diagnosed with COVID-19 but who did not have SCD/SCT. After 1:1 propensity score matching (based on age, sex, and other preexisting comorbidities), patients with COVID-19 and SCD remained at a higher risk of hospitalization (relative risk [RR], 2.0; 95% CI, 1.5-2.7) and development of pneumonia (RR, 2.4; 95% CI, 1.6-3.4) and pain (RR, 3.4; 95% CI, 2.5-4.8) compared with Black persons without SCD/SCT. The case fatality rates for those with SCD compared with Black persons without SCD/SCT were not significantly different. There also were no significant differences in COVID-19 outcomes between individuals with SCT and Black persons without SCD/SCT within the matched cohorts. Copyright © 2021 by The American Society of Hematology.

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1. **COVID-19 pneumonia in a pediatric sickle cell patient requiring red blood cell exchange**  
   Walker Shannon C. Clinical case reports 2021;9:1367-1370.

Patients with sickle cell disease are already at high risk for respiratory complications, which SARS-CoV-2 can rapidly worsen. The case emphasizes the importance of efficiently maximizing standard therapies in sickle cell patients with COVID-19. Copyright © 2021 The Authors. Clinical Case Reports published by John Wiley & Sons Ltd.

[Available online at this link](https://www.knowledgeshare.nhs.uk/index.php?PageID=link_resolver&link=cd863a23c892d88a36b267ed5d84e4d4)

1. **Covid-19 presentation in patients with sickle cell disease: A case series**  
   Chen-Goodspeed A. Journal of Investigative Medicine 2021;69:551.

Purpose of Study Severe Acute Respiratory Distress Syndrome Coronavirus 2, also known as COVID-19, has been shown to cause adverse health effects to those with comorbidities, one of which is sickle cell disease (SCD). By examining cases of SCD patients with COVID-19, we aim to examine the impact of COVID-19 on SCD patients and their clinical presentations. Methods Used In this case series, we will report on five SCD patients who presented with symptoms ranging in severity from mildly symptomatic to weeklong hospitalizations. Summary of Results The firstcase is a 30-year-old man with HbSS who was hospitalized for four days with vaso-occlusive crisis (VOC) and hypoxia due to COVID-19 infection. Case two is a 30-year old man with HbSS who was hospitalized for two days after presenting to the ED for VOC and COVID-19. Case three is a 49-year-old woman with HbSC who presented to the ED with VOC and COVID-19. The fourth case is a 23-year-old man with HbSS who was hospitalized for eight days after he presented with VOC, acute chest syndrome, and COVID-19. The final case is a 25-year-old woman with HbSS who presented with cough and loss of taste who had mild symptoms. Conclusions These cases show that the presentation of COVID-19 in SCD patients is not always the typical COVID- 19 triad of cough, fever, and dyspnea that clinicians recognize. Recent case reports on SCD patients have indicated that COVID-19 can trigger VOC in SCD patients by increasing inflammatory cytokines such as interleukin-6. Thus patients with SCD and COVID-19 can present with a VOC rather than typical COVID-19 symptoms, as seen in four of these cases. Moreover, as seen in this series, the findings on chest radiologic imaging for this subset of patients can also differ from the typical presentation of the COVID-19 chest radiologic findings. In conclusion, we recommend that SCD patients with COVID-19 exposure or who present with VOC, even in the absence of the typical signs and symptoms of COVID-19 infection, be tested for COVID-19. Individuals, especially adults, with SCD commonly have preexisting multi-organ dysfunction and delicate immune system which put them at increased risk for COVID-19 infection-related morbidity and mortality particularly when this infection is unrecognized and improperly treated.

[Available online at this link](https://www.knowledgeshare.nhs.uk/index.php?PageID=link_resolver&link=ba708e573ae6c3d5d16e2b7988e3cfc3)

1. **Estimated SARS-CoV-2 Seroprevalence in Healthy Children and Those with Chronic Illnesses in The Washington Metropolitan Area as of October 2020**  
   Bahar Burak 2021;:No page numbers.

The estimated SARS-CoV-2 seroprevalence in children was found to be 9.46% for the Washington Metropolitan area. Hispanic/Latinx individuals were found to have higher odds of seropositivity. While chronic medical conditions were not associated with having antibodies, previous fever and body aches were predictive symptoms.

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1. **Guideline on the peri-operative management of patients with sickle cell disease: Guideline from the Association of Anaesthetists**  
   Walker I. Anaesthesia 2021;:No page numbers.

Sickle cell disease is a multisystem disease characterised by chronic haemolytic anaemia, painful vaso-occlusive crises and acute and chronic end-organ damage. It is one of the most common serious inherited single gene conditions worldwide and has a major impact on the health of affected individuals. Peri-operative complications are higher in patients with sickle cell disease compared with the general population and may be sickle or non-sickle-related. Complications may be reduced by meticulous peri-operative care and transfusion, but unnecessary transfusion should be avoided, particularly to reduce the risk of allo-immunisation. Planned surgery and anaesthesia for patients with sickle cell disease should ideally be undertaken in centres with experience in caring for these patients. In an emergency, advice should be sought from specialists with experience in sickle cell disease through the haemoglobinopathy network arrangements. Emerging data suggest that patients with sickle cell disease are at increased risk of COVID-19 infection but may have a relatively mild clinical course. Outcomes are determined by pre-existing comorbidities, as for the general population. Copyright © 2021 The Authors. Anaesthesia published by John Wiley & Sons Ltd on behalf of Association of Anaesthetists.

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1. **Is sickle cell disease a risk factor for severe COVID-19 outcomes in hospitalized patients? A multicenter national retrospective cohort study**  
   Abdulrahman Abdulkarim EJHaem 2021;:No page numbers.

Introduction: Studies that examine the association between sickle cell disease (SCD) and COVID-19 outcomes are lacking. This study aims to determine whether SCD is a risk factor for severe COVID-19 infection in regard to the requirement of noninvasive ventilation/high flow nasal cannula (NIV/HFNC), mechanical ventilation (MV), or death in hospitalized patients., Methods: Retrospective cohort study included COVID-19 patients admitted to four COVID-19 treatment facilities in Bahrain between February 24, 2020 and July 31, 2020. All SCD patients with COVID-19 were included and compared to a randomly selected sample of non-SCD patients with COVID-19. Data were collected from the medical records. Multivariate logistic regression models were used to control for confounders and estimate the effect of SCD on the outcomes., Results: 1792 patients with COVID-19 were included; 38 of whom were diagnosed with SCD as well. In the SCD group, one (2.6%) patient required NIV/HFNC, one (2.6%) required MV, and one (2.6%) death occurred. In comparison, 56 (3.2%) of the non-SCD patients required NIV/HFNC, 47 (2.7%) required MV, and death occurred in 58 (3.3%) patients. Upon adjusting for confounders, SCD had an odds ratio of 1.847 (95% CI: 0.39-8.83; p = 0.442)., Conclusion: Our results indicate that SCD is not a risk factor for worse COVID-19 outcomes in hospitalized patients. Copyright © 2021 The Authors. eJHaem published by British Society for Haematology and John Wiley & Sons Ltd.

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1. **Rapid development of seizures and PRES in a COVID-19 patient**  
   Santos de Lima Fabiane Epilepsy & behavior reports 2021;15:100436.

Neurological dysfunction has been noted in up to 36% of patients hospitalized with COVID-19, and a variety of mechanisms of neurological injury are possible. Here we report the rapid development of PRES and acute seizures in a patient with COVID-19 infection and sickle cell disease. The combination of COVID and sickle cell disease may raise the risk of PRES and could contribute to the higher mortality rate of COVID in patients with sickle cell disease. Copyright © 2021 The Author(s).

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1. **Sickle Cell Anemia Presenting with Vaso-Occlusive Pain: Should We Screen for COVID-19?**  
   Ali M. Dubai Medical Journal 2021;:No page numbers.

Despite the widespread of coronavirus disease-19 (CO-VID-19) infection around the world, there are very scarce reported literature about the care of patients with a known diagnosis of hemoglobin disorders such as sickle cell disease (SCD) or thalassemia and confirmed COVID-19 infection. Thalassemia International Federation issued a position statement to include patients with thalassemia and SCD among the high-risk groups of patients. Here, we present an interesting case of a 42-year-old patient know to have SCD presenting with Vaso-occlusive (VOC) pain episode in the absence of COVID-19 signs and symptoms, who tested positive for COVID-19 infection and had a smooth recovery. This case highlights the importance of screening SCD patients presenting with VOC-related events even in the absence of COVID-19 signs and symptoms. Copyright © 2021 The Author(s). Published by S. Karger AG, Basel.

[Available online at this link](https://www.knowledgeshare.nhs.uk/index.php?PageID=link_resolver&link=66acf611a1feaa16938bdcc409b97642)

1. **Stroke presentation in an adolescent with mild hemoglobin scdisease**  
   Bennett J. M. Pediatrics 2021;147:1056.

Background: In individuals affected with Hemoglobin SC (HbSC) disease, one beta globin gene is affected by thesickle mutation (position 6, a single-base pair change encodes valine instead of glutamine) while the other betaglobin gene contains a mutation for hemoglobin C (position 6, lysine is encoded instead of glutamine). In thiscompound heterozygous state individuals typically present with a milder sickle cell disease (SCD) coursecompared to those with homozygous HbS (HbSS) disease. Although children with HbSC disease experience asignificant incidence of silent cerebral infarcts, acute stroke presentation is exceptionally rare. Herein wedescribe a case of an adolescent male with historically uncomplicated HbSC disease, and otherwiseunremarkable sickle cell surveillance labs on admission presenting with new acute onset stroke. CasePresentation: Our patient is a 16 year old male with uncomplicated HbSC disease and no history ofhospitalizations for vasoocclusive crisis. He presented with 48 hours of difficulty focusing out of his left eye.Physical exam revealed impaired right eye adduction, left eye nystagmus on lateral left gaze, and decreased sensation to touch to the left hemi face and left arm. Strength and deep tendon reflexes were normal. Normalgait was observed and he had no dysdiadochokinesia. STAT MRI orbit and brain with contrast revealedmultiple foci of acute infarct involving the pons and left cerebral peduncle. Vessel imaging with MRA wasconcerning for internuclear ophthalmoplegia secondary to stenosis in the posterior cerebral circulation. Vitalsigns were normal for age and labs were overall mild and unchanged from his baseline (WBC 8750/mcL,Hemoglobin 13.5g/dL, Platelets 290,000/mcL, Reticulocyte 2.11%, CRP <0.30mg/dL). Initial thrombophilia workup demonstrated no increase in antiphospholipid antibodies, normal homocysteine levels and fibrinogenlevels. Fibrin D-dimer was mildly elevated at 0.55mcg/mL. Further thrombophilia evaluation is ongoing. Arespiratory viral panel and COVID-19 testing were performed and were negative. The patient was treated withemergent complete exchange transfusion. Discussion(s): This case highlights the importance recognizing the riskfor acute stroke in patients with HbSC. Prior to this acute presentation of stroke, this patient had mild disease with no history of prior transfusions and mild vasooclussive crises managed as an outpatient. Earlyrecognition, imaging and management for those who present with focal neurologic deficits is essential inpreventing considerable morbidity and mortality. It is imperative that as a general pediatrician, one must bevigilant and not rely on reassuring labs or prior histories of mild disease presentations in patients with SCD.Unlike with HbSS, there is limited data and consensus guidelines for secondary stroke prevention in patients with HbSC, thus emphazing clinical judgement in initiation of exchange transfusion. We did proceed withexchange transfusion taking into consideration the data that supports improved outcomes in HbSS patients.

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1. **Association of Sickle Cell Trait with Risk and Mortality of COVID-19: Results from the UK Biobank**  
   Resurreccion Kyle 2020;:No page numbers.

We tested the hypothesis that patients with sickle cell trait (SCT), a common condition in individuals of African descent, have increased risk and mortality for coronavirus disease (COVID-19) in the UK Biobank. By June 17, 2020, 1,550 of 7,668 (20%) tested subjects were positive for COVID-19, including 298 (19%) deaths. Blacks had higher rates than Whites for COVID-19 infections (79/222=36% vs. 1,342/7,010=19%, P =1.28×10 −9 ). Among Blacks, SCT carriers did not have higher infection rates (5/15=33%) than non-SCT carriers (74/207=36%), P =1.00. However, SCT carriers had a trend of higher death rates (2/5=40%) than non-SCT carriers (12/74=16%), although not statistically significant ( P =0.21).

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1. **Blood flow, capillary transit times, and tissue oxygenation: the centennial of capillary recruitment**  
   Ostergaard Leif Journal of applied physiology (Bethesda, Md. : 1985) 2020;129:1413-1421.

The transport of oxygen between blood and tissue is limited by blood's capillary transit time, understood as the time available for diffusion exchange before blood returns to the heart. If all capillaries contribute equally to tissue oxygenation at all times, this physical limitation would render vasodilation and increased blood flow insufficient means to meet increased metabolic demands in the heart, muscle, and other organs. In 1920, Danish physiologist August Krogh was awarded the Nobel Prize in Physiology or Medicine for his mathematical and quantitative, experimental demonstration of a solution to this conceptual problem: capillary recruitment, the active opening of previously closed capillaries to meet metabolic demands. Today, capillary recruitment is still mentioned in textbooks. When we suspect symptoms might represent hypoxia of a vascular origin, however, we search for relevant, flow-limiting conditions in our patients and rarely ascribe hypoxia or hypoxemia to short capillary transit times. This review describes how natural changes in capillary transit-time heterogeneity (CTH) and capillary hematocrit (HCT) across open capillaries during blood flow increases can account for a match of oxygen availability to metabolic demands in normal tissue. CTH and HCT depend on a number of factors: on blood properties, including plasma viscosity, the number, size, and deformability of blood cells, and blood cell interactions with capillary endothelium; on anatomical factors including glycocalyx, endothelial cells, basement membrane, and pericytes that affect the capillary diameter; and on any external compression. The review describes how risk factor- and disease-related changes in CTH and HCT interfere with flow-metabolism coupling and tissue oxygenation and discusses whether such capillary dysfunction contributes to vascular disease pathology.

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1. **Can Increased Recovery Rates from Coronavirus be explained by Prevalence of ADHD? An Analysis at the US Statewide Level**  
   Arbel Yuval Journal of attention disorders 2020;:1087054720959707.

Previous research demonstrates that ADHD is considered a risk factor for COVID-19. The current study attempts to investigate the relationships between infection, mortality and recovery rates from coronavirus and the prevalence of ADHD at the US statewide level. Based on information from 2011 regarding the prevalence of ADHD across the US by state, findings suggest that, while there are no correlations between ADHD and population size, infection and mortality rates from coronavirus, recovery rates (recovery-population ratio) rise with the prevalence of ADHD. Consequently, a possible explanation is that in coping with the disease, ADHD might provide an evolutionary advantage. An example of this phenomenon can be found in the gene that causes sickle-cell disease, which, as a non-dominant gene, helps cope with infection from malaria. If corroborated, research findings may support the conclusion that coronavirus limitations in special educational frameworks for ADHD would not be required or could be relaxed.JEL Codes: H75, I12.

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1. **Caring for patients with sickle cell disease during a pandemic: Continuing to provide automated red blood cell exchange transfusions in difficult times**  
   Godby R. C. Blood 2020;136:25-26.

Split-Screen Share V Tools V Introduction: The World Health Organization declared COVID-19 a global pandemic on 03/11/20. Subsequent concerns around caring for patients with sickle cell disease who require automated red blood cell (RBC) exchange transfusions emerged, especially in the setting of physical distancing and national shortages in blood product supplies. In this vulnerable population at high risk of alloimmunization, ideal transfusion parameters (e.g., antigen optimization) will likely grow increasingly difficult to satisfy and require careful evaluation and strategic planning. Method(s): Automated RBC exchange transfusions were performed at the University of Alabama at Birmingham (UAB) in patients with sickle cell disease for a variety of clinical indications with the SprimaryobiectJVfeof lowering the amount of Hemoglobin S (goal 15%) and replacing it with Hemoglobin A. We collected the number of weekly RBC exchange transfusions performed and then compared the frequencies between 01/05/20 and 03/14/20 (pre-pandemic) to those between 03/15/20 and 08/01/20 (intra-pandemic) using a one-tailed t-test. We also examined the number of RBC units ordered per week at UAB, in both the inpatient and outpatient settings, shortly before and after the declaration of a global pandemic using a one-tailed t-test. Result(s): The mean frequency of RBC exchange transfusions performed per week was 8.1 [standard deviation 2.3] pre-pandemic and 8.6 [2.3] intra-pandemic (Figure 1a). There was no statistically significant difference (p=0.27) in the frequency between these two periods. Shortly prior to the start of the pandemic (02/23/20-03/14/20), a mean of 77.3 [17.9] units/week were ordered for outpatient RBC exchange transfusions. Shortly after the start of the pandemic (03/15/20-04/26/20), a mean of 55.3 [22.8] units/week were ordered for outpatient RBC exchange transfusions, which was also not significantly different (p=0.09). During this time period, the mean number of RBC units per week ordered in the inpatient surgical setting significantly declined from 719.3 [43.1] to 390.0 [46.8] as elective procedures were delayed (p<0.005) (Figure 1b). Conclusions/Future Directions: The frequency of automated RBC exchange transfusions performed at UAB did not decrease after the onset of the pandemic. UAB was able to continue caring for patients with sickle cell disease receiving RBC exchange transfusions as the pandemic emerged and national blood product supplies declined despite a similar overall demand. Interestingly, there was also a concomitant decrease in the demand for RBCs from inpatient surgical settings as elective procedures were delayed, possibly contributing to the blood bank's ability to maintain ideal transfusion parameters and perform antigen optimization of transfused RBCs. As the COVID-19 pandemic continues, the national shortage of blood product supplies will likely worsen and necessitate multidisciplinary efforts, including intra-institutional and inter-institutional collaborations, to continue caring for patients with sickle cell disease receiving RBC exchange transfusions. Furthermore, community education, safely structured blood drives, and other efforts to encourage donations are essential to maintain the national blood product supply.

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1. **Challenges in the Management of Sickle Cell Disease During SARS-CoV-2 Pandemic**  
   Alsayegh Faisal Clinical and applied thrombosis/hemostasis : official journal of the International Academy of Clinical and Applied Thrombosis/Hemostasis 2020;26:1076029620955240.

The management of sickle cell disease (SCD) and its complications in the COVID-19 era is very challenging. The recurrent sickling process in SCD causes tissue hypoxemia and micro-infarcts, resulting in end organ damage. Since the outbreak of SARS-CoV-2 pandemic, little data has been published about SCD concerning clinical presentation with COVID-19 and management. Hydroxyurea has been the cornerstone of management in children and adults with SCD, with evidence of its effect on controlling end organ damage. There are several anti-sickling drugs that have been approved recently that might have an additive value toward the management of SCD and its complications. The role of simple and exchange transfusions is well established and should always be considered in the management of various complications. The value of convalescent plasma has been demonstrated in small case series, but large randomized controlled studies are still awaited. Immunomodulatory agents may play a role in reducing the damaging effects of cytokines storm that contributes to the morbidity and mortality in advanced cases. Prophylactic anticoagulation should be considered in every management protocol because SCD and COVID-19 are thrombogenic conditions. Management proposals of different presentations of patients with SCD and COVID-19 are outlined.

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1. **Characteristics and risk factors associated with critical illness in pediatric COVID-19**  
   Fisler Grace Annals of intensive care 2020;10:171.

BACKGROUND: While much has been reported regarding the clinical course of COVID-19 in children, little is known regarding factors associated with organ dysfunction in pediatric COVID-19. We describe critical illness in pediatric patients with active COVID-19 and identify factors associated with PICU admission and organ dysfunction. This is a retrospective chart review of 77 pediatric patients age 1 day to 21 years admitted to two New York City pediatric hospitals within the Northwell Health system between February 1 and April 24, 2020 with PCR + SARS-CoV-2. Descriptive statistics were used to describe the hospital course and laboratory results and bivariate comparisons were performed on variables to determine differences., RESULTS: Forty-seven patients (61%) were admitted to the general pediatric floor and thirty (39%) to the PICU. The majority (97%, n = 75) survived to discharge, 1.3% (n = 1) remain admitted, and 1.3% (n = 1) died. Common indications for PICU admission included hypoxia (50%), hemodynamic instability (20%), diabetic ketoacidosis (6.7%), mediastinal mass (6.7%), apnea (6.7%), acute chest syndrome in sickle cell disease (6.7%), and cardiac dysfunction (6.7%). Of PICU patients, 46.7% experienced any significant organ dysfunction (pSOFA > = 2) during admission. Patients aged 12 years or greater were more likely to be admitted to a PICU compared to younger patients (p = 0.015). Presence of an underlying comorbidity was not associated with need for PICU admission (p = 0.227) or organ dysfunction (p = 0.87). Initial white blood cell count (WBC), platelet count, and ferritin were not associated with need for PICU admission. Initial C-reactive protein was associated with both need for PICU admission (p = 0.005) and presence of organ dysfunction (p = 0.001). Initial WBC and presenting thrombocytopenia were associated with organ dysfunction (p = 0.034 and p = 0.003, respectively)., CONCLUSIONS: Age over 12 years and initial CRP were associated with need for PICU admission in COVID-19. Organ dysfunction was associated with elevated admission CRP, elevated WBC, and thrombocytopenia. These factors may be useful in determining risk for critical illness and organ dysfunction in pediatric COVID-19.

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1. **Children and young adults admitted to a NYC children's hospital had a similar rate of severe COVID-19 coagulopathy as that reported in older adults**  
   Mitchell W. B. Blood 2020;136:27-28.

Split-Screen Share v Tools v The coagulopathy associated with COVID-19 has not been previously described in children and young adults. We reviewed the clinical and laboratory characteristics of children and young adults admitted for COVID-19 to an urban Children's Hospital in New York City, focusing on coagulation and venous thromboembolism. Clinical and laboratory data were analyzed from 54 patients aged 2 months to 30 years treated by the Pediatric Hematology service at a single Children's Hospital between January 1 and May 31, 2020. Information was obtained from hospital records with IRB approval. There was a moderate male predominance, with 32 (59%) males and 22 (41%) females. There were 28 (52%) patients younger than 18 years and 26 (48%) patients 18 - 30 years old. 26% of patients identified as Black, and 57% as Hispanic/Latino, similar to the community demographics in the Bronx in the 2019 census. Obesity was the most prevalent comorbid condition, with 19 (35%) patients having BMI of 30 or higher. There were also 12 (22%) patients with sickle cell anemia. There were 28 (52%) patients in this cohort with severe and critical illness, as based on established criteria, and 25 (46%) patients required increased ventilatory support. This was defined by the need for > 5L nasal cannula, high-flow nasal cannula, non-rebreather, or intubation. 11 patients (20%) had documented venous thromboembolism (VTE). Four patients died of COVID-19 complications at ages 2 months, 11, 14 and 18 years old. The VTE rate was similar in those patients under 18 years of age (5 of 28, 18%) and those 18 - 30 years of age (6 of 26, 23%). Most (94%) patients had a D-Dimer > 0.5 (upper limit of normal) at admission and 57% developed peak D-Dimer > 5 ug/mL during their admission. Elevated D-dimer > 5 was a risk factor for VTE with 3 of 23 (13%) and 7 of 17 (41%) patients developing VTE with D-dimer < 5 and > 5, respectively (OR 4.7, p=0.042). Patients requiring increased ventilatory support had a 36% rate of VTE as compared to 1 of 28 (4%) of those without (OR 15.2, p=0.003). Six of 24 patients on prophylactic anticoagulation developed VTE. One patient developed a pulmonary embolism 10 days post discharge from the hospital. No patients on anti-Xabased low molecular weight heparin prophylaxis developed VTE. None of 12 patients with sickle cell anemia developed VTE, had peak D-Dimer > 5 ug/mL or required increased ventilatory support. Hospitalized children and young adults with COVID-19 in our cohort developed a coagulopathy similar to that of older adults, characterized by elevated D-Dimer and high rate of VTE. This is in contrast to the published pediatric series out of China and Singapore that described mild illness and did not comment on VTE rates. Presence of elevated D-dimer or need for increased ventilatory support were significant risk factors for thrombosis. Patients with sickle cell anemia had a lower risk of VTE and less severe illness. Anti-Xa monitored thromboprophylaxis may aid in preventing or ameliorating the COVID-19 coagulopathy in children and young adults. Institutional anticoagulation guidelines were developed based on these observations. (Table Presented).

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1. **Clinical course and outcomes of sickle cell disease patients withCOVID-19 at a new york hospital**  
   Mazloom A. Blood 2020;136:34-36.

Introduction: The outbreak of a novel infection, COVID-19, has greatly impacted the well-being of individualsworldwide. Persons with sickle cell disease (SCD) constitute a vulnerable population, subject tohealth disparities, who may have worse outcomes from COVID-19. Within the United States, NewYork has a large population of patients with SCD. Here, we analyze the clinical course and outcomesof SCD patients with COVID-19 who were admitted to a community teaching hospital in Brooklyn,NY. Methods : We conducted a retrospective chart review of adult patients with SCD hospitalized with laboratory-confirmed COVID-19. Electronic health records were reviewed to identify patients and analyze theirclinical course. Clinical characteristics, laboratory and radiology data were assessed. Rates of acutechest syndrome (ACS), acute kidney injury (AKI) and venous thromboembolism (VTE) weredetermined. ACS was defined by the presence of fever and/or respiratory symptoms accompaniedby a new pulmonary infiltrate on chest Xray. Data on use of blood transfusion, treatments, length ofstay and mortality were collected. Results : Between March 1 to June 30, 2020, 53 adults with SCD were hospitalized at our institution. Of these,13 patients had COVID-19 infection. The mean (+/-SD) age of the COVID-19 patients was 34+/-10 years(range, 22 to 50) with 54% being female. Seven patients (54%) were Hb SS, and 6 patients (46%)were Hb SC. Comorbid conditions included Diabetes Mellitus (1 patient), SLE (1), End-stage renaldisease (1), prior VTE (4) and Avascular necrosis of hip (3). Four patients were on hydroxyurea. Clinical, laboratory and radiological findings are summarized in Table 1. While all the Hb SS patientspresented with vaso-occlusive crisis, 4 of the 6 patients with Hb SC did not have symptoms of paincrisis. Chest pain and cough were the most common symptoms at presentation. During the hospitalstay, 12 patients (92%) had at least one febrile episode >38degreeC, with 77% having recurrent feversabove 38.5degreeC. Eleven patients (85%) met criteria for ACS. Seventy-seven percent of all patients requiredsupplemental oxygen. Nine patients (69%) were transfused, with 4 patients undergoing exchangetransfusion. Sixty-seven percent of the transfused patients were transfused within 48 hours ofadmission. No patients required intubation or mechanical ventilation and none were admitted to theintensive care unit (ICU). Five patients (38.5%) received hydroxychloroquine while 84.6% weretreated with antibiotics. No patient received remdesivir. Three patients (23%) developed AKI: of these, one patient required acute hemodialysis, the other twocases were mild with peak creatinine less than 2.0 mg/dl. Ninety-two percent of patients received prophylactic anticoagulation with either unfractionatedheparin, enoxaparin or fondaparinux. One patient who did not receive an anticoagulant due tothrombocytopenia developed an acute deep vein thrombosis which was also catheter-related. Ofnote, during the initial phase of the pandemic standard dosing of prophylactic anticoagulants wereused but in the later months, some patients received higher prophylactic doses in keeping withhospital protocol. The median length of hospital stay was 9.4 days (interquartile range, 8.1 to 13.3). There were nodeaths - all patients were discharged home. Summary: Panepinto et al (Emerg Infect Dis.) reported a mortality of 7% in 178 SCD patients with COVID-19 inthe United States. Other published reports have detailed more favorable outcomes (Arlet et al,Lancet and Appiah-Kubi et al, Br J Haematol.). In this small retrospective analysis of hospitalizedSCD patients, there was no mortality. Acute chest syndrome was the most common complication observed. VTE and severe AKI were infrequent. Blood transfusion was performed in the majority ofpatients (69%); two thirds of the patients transfused received blood within 48 hours ofhospitalization. There were no ICU admissions and no use of mechanical ventilation indicativeperhaps of less severe COVID-19 disease. This may have been due to the young age o the cohort.Early use of blood transfusion may have been a factor in reducing disease severity and improvingoutcomes. The best approach to managing these patients is unclear. We advocate for thedevelopment and dissemination of evidence-based guidelines to manage SCD patients with COVID-19 to reduce morbidity and mortality in this at-risk population.

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1. **Clinical Spectrum of Sickle Cell Disease and COVID-19: Laboratory and Clinical Factors Associated with Morbidity and Mortality**  
   Minniti Caterina 2020;:No page numbers.

Background: The COVID-19 pandemic has generated concern as a potential remarkably severe threat to the sickle cell disease (SCD) population. We aim to identify predictors of outcomes and survival in a large US-based SCD and COVID-19 cohort to inform best approaches to prevention and care.&lt;br&gt;&lt;br&gt;Methods: Clinical data were collected at baseline and during the clinical course using a standardized form in SCD patients diagnosed with COVID-19 at 5 academic centers in four COVID-19 epicenters. Patients were followed post-hospital discharge for up to 3 months.&lt;br&gt;&lt;br&gt;Results: Of 66 consecutive SCD patients with COVID-19, 75% required hospitalization, with a median length of stay of 6 days, and 7 died (10.6%). Patients with preexisting kidney disease were more likely to be hospitalized, while age, sex and genotype had no effect. The most common presenting symptom was vaso-occlusive pain. Chest X-ray was abnormal (acute chest syndrome) at presentation in 30 of 50 (61%) hospitalized and all deceased patients. Older age (median of 53 versus 32 years) and histories of pulmonary hypertension, congestive heart failure and/or stroke were more prevalent in deceased patients, as were high creatinine, lactate dehydrogenase, C-reactive protein, and D-dimers. The use of anti-coagulation, but not hydroxychloroquine or transfusion, during inpatient hospitalization was associated with decreased mortality (p&amp;lt;0.05). All deaths occurred in individuals not taking hydroxyurea or other SCD modifying therapy.&lt;br&gt;&lt;br&gt;Conclusions: Patients with SCD and COVID-19 infection demonstrated a broad range of disease severity, from mild to very severe. COVID-19 in SCD individuals with pre-existing cardiopulmonary, renal disease and/or stroke presenting with pain and high creatinine should be considered at risk of death, irrespective of genotype or gender. Inpatient use of anticoagulation should be considered for all SCD patients with COVID-19. Though older individuals with vasculopathic comorbidities and high D-dimers were more likely to die, the median age of death was decades lower than the non-SCD population.&lt;br&gt;&lt;br&gt;Funding Statement: None.&lt;br&gt;&lt;br&gt;Declaration of Interests: None&lt;br&gt;&lt;br&gt;Ethics Approval Statement: Each respective Institutional Review Board approved data collection as minimal-risk research and waived the requirement for informed consent.

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1. **Coagulation Status and Venous Thromboembolism Risk in African Americans: A Potential Risk Factor in COVID-19**  
   Frydman Galit H. Clinical and applied thrombosis/hemostasis : official journal of the International Academy of Clinical and Applied Thrombosis/Hemostasis 2020;26:1076029620943671.

Severe acute respiratory syndrome coronavirus 2 infection (COVID-19) is known to induce severe inflammation and activation of the coagulation system, resulting in a prothrombotic state. Although inflammatory conditions and organ-specific diseases have been shown to be strong determinants of morbidity and mortality in patients with COVID-19, it is unclear whether preexisting differences in coagulation impact the severity of COVID-19. African Americans have higher rates of COVID-19 infection and disease-related morbidity and mortality. Moreover, African Americans are known to be at a higher risk for thrombotic events due to both biological and socioeconomic factors. In this review, we explore whether differences in baseline coagulation status and medical management of coagulation play an important role in COVID-19 disease severity and contribute to racial disparity trends within COVID-19.

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1. **Combination dose-escalated hydroxyurea and transfusion: an approach to conserve blood during the COVID-19 pandemic**  
   Nickel Robert Sheppard Blood 2020;135:2320-2322.

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1. **COVID 19 and hemoglobinopathies: update of the Italian experience**  
   Motta I. Blood 2020;136:17-18.

Background. Patients with pre-existent chronic morbidities are likely to be more severely affected by SARS-Cov2 infection. In Italy, the "Societa Italiana Talassemie ed Emoglobinopatie" (SITE) has recently estimated the number of patients (Pts) with Hemoglobinopathies followed by Italian Specialized Centers (SITE Network). Five thousand Transfusion-dependent beta-thalassemia (TDT), 1900 Non-Transfusion-dependent beta-thalassemia (NTDT) and 2000 Sickle Cell Disease (SCD) were registered [1]. To verify the impact of SARS-CoV-2 infection on Pts with Hemoglobinopathies, we performed a specific survey by electronic Case Report Form (eCRF). Inclusion criteria included positive swab or serology in a patient with hemoglobinopathy and at least 15 days of follow-up from either the onset of symptoms or SARS-CoV2 positivity. The survey was approved by the Ethics Committee, and eCRF was shared with the Centers of Italian Hemoglobinopathies Network. Preliminary data updated to April 10, 2020, were published [2]. Results. As of July 31, 2020, 27 cases have been reported: 18 TDT, 4 NTDT, 5 SCD. 89% of the cases were in Northern Italy, where the rate of infection was much higher than the rest of the country, reflecting the national epidemiology. The mean age of thalassemia patients (TDT and NTDT) was 43+/-11 years, and 55% were male; the mean age of SCD patients was 33+/-15 years, and 40% was male. The likely source of infection has been detected in 63% (17/27) of cases: 11 had occupational exposure, 6 had a positive relative. Five patients were asymptomatic: for them, the SARS-CoV-2 infection was identified by positive swab for 1 patient and by positive level of IgG for 4. Twenty patients had associated comorbidities, 14 were splenectomized, and 3 had functional asplenia. Eleven patients were hospitalized, only one in high-intensity care unit. Three patients required more intensive ventilation support with continuous positive airway pressure (CPAP), one of these has a history of diffuse large B-cell lymphoma treated with chemotherapy in the previous year. Three other patients required support by oxygen. No Pts required intubation. Two Pts increased blood requirement. Only five received supposedly specific treatment for COVID-19: two hydroxychloroquine (HCQ), one HCQ plus ritonavir/darunavir, and one HCQ plus anakinra, one HCQ plus Tocilizumab plus Lopinavir/Ritonavir. The clinical course of hospitalized patients was 18+/-7 days. All patients recovered. Conclusions. The prevalence of COVID-19 infection in Italian patients with Hemoglobinopathies result 0,3% while in general population the prevalence in Italy is 0,4% [3]. Considering that the thalassemia population is more strictly observed, we could postulate that the precautions suggested or self-applied by the Pts were effective. No death nor severe SARS with intubation, nor signs of cytokines storm, only one thromboembolic event was observed although most individuals had preexisting complications. A single case with pulmonary hypertension has been described in detail [4]. In most individuals the infection has been pauci or asymptomatic and all recovered. This experience differs from what has been observed in Iran on a similar series with different severity and mortality and ask for a more in-depth comparison [5]. In conclusion, our data do not indicate increased severity of COVID-19 in Pts with Hemoglobinopathies followed in Specialized Centers.

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1. **Covid delays in diagnosis and treatment of early onset crc**  
   Lee-Allen J. American Journal of Gastroenterology 2020;115:S848-S849.

INTRODUCTION: This is a case of early onset Colorectal cancer (CRC) in which the COVID pandemic caused both diagnosis and treatment delays. CASE DESCRIPTION/METHODS: A 31-year-old male with controlled Sickle Cell Anemia (no crises in over 10 years) presented to Emergency Department with 6 months of non-focal abdominal discomfort, 30lb weight loss and CT one month prior with 5 cm, Ascending Colon lesion [Img. 1]. Chief compliant was an expedited colonoscopy as COVID policies eliminated all outpatient and nonemergent case requests. Labs were notable for hemoglobin 9.6 g/dL (down-trending since 2019, from baseline of 11-13 g/dL) and MCV 59 FL. He denied upper gastrointestinal symptoms, family history of CRC or overt gastrointestinal bleeding. He tested negative for COVID. Outpatient colonoscopy, 4 days later, revealed a circumferential, ulcerated, firm ascending colon mass with malignant appearance [Img. 2]. Same day CT chest, abdomen and pelvis were negative for metastases. Pre-surgical COVID testing returned positive. Surgery was postponed until two negative COVID tests. Ascending colon biopsies confirmed invasive, moderately, differentiated adenocarcinoma, CK 20 and CDX-2 positive. Two weeks later, his COVID criteria was met and right hemi-colectomy was completed (tumor size: 8 x 5 x 1cm). Pathology confirmed invasive adenocarcinoma without any lymph nodes and negative margins. Stains were negative for hereditary syndromes. The patient did well after surgery. DISCUSSION: This patient's immediate early onset CRC outcome was not complicated by COVID delays. Research support delaying surgical intervention in cancer patients whether COVID negative or positive due to high risk for COVID specific mortality [1]. Future research is needed to determine the 5-10-year impact of COVID delays on CRC detection, stage, treatment and mortality. Further, prior studies have shown that colonoscopy delays of greater than 10 months from time of positive FIT led to increased detection of advanced stage CRC compared to colonoscopy within 3-6 months [2]. Thus, in the next 3-6 months Gastroenterologists should prioritize outreach and completion of colonoscopies for patients suspected of malignancy in the pre-COVID era. (Figure Presented).

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1. **COVID symptoms and COVID anxiety in sickle cell disease**  
   Smith W. R. Blood 2020;136:16-17.

Background Even before the US upswing of the current COVID pandemic, the number of sickle cell disease (SCD)patients coming to hospitals and EDs appeared to fall drastically. This happened despite SCDpatients having often been heavy utilizers of the ED and hospital for their iconic vaso-occlusive crises(VOC). Though ambulatory SCD clinics quick converted largely to telehealth in order to comply withstay-at-home orders designed to suppress person-to-person transmission, some SCD patientsappeared to avoid care, delay care, or refuse doctors' invitations for care. Presumably patients did soout of COVID fears, but this has not been confirmed in the literature. Further, whether these patientshad COVID symptoms but stayed at home has not been studied. As part of quality improvement (QI)to conduct COVID surveillance in an adult sickle cell program, we sought to explain and predict SCDhealth care utilization patterns we were observing, as well as to determine urgent physical and mental health needs of patients who appeared to be avoiding care. Methods Fifteen sta in the Adult Sickle Cell Medical Home at Virginia Commonwealth University, a largeurban academic medical center, conducted a telephone survey ("wellness check"was used when wetalked to patients) of all known adults with SCD over 19 days in 2020. A sta member confirmed thepatient had SCD, asked permission to proceed, then asked about symptoms consistent with COVID-19. At the end of the telephone survey, respondents wer invited to complete an email survey of sicklecell and COVID-19 utilization attitudes (19-33 items, depending on the response pattern, either drawnfrom the National Health Interview Survey, from the Adult Sickle Cell Quality of Life Measurementquality of care survey, or drafted by the authors), the Sickle Cell Stress Survey-Adult (SCSS-A, a 10-item previously validated survey), and anxiety and depression (PHQ9 of the PRIME-MD). Results Of 622 adults approached by phone call, 353 responded to the following yes/no screening questionsregarding the prior 14 days: fever over 100 F 0/353 (0.00%); cough 3/353(0.01%); diiculty breathing0/353(0.00%); unexplained shortness of breath 2/353(0.01%); sore throat 2/353 (0.01%); unexplainedmuscle soreness 2/353(0.01%);contact with anyone who tested positive for COVID-19 2/353(0.01%);testing for COVID 19 6/353(0.02%). For QI purposes, we set a threshold of three or more COVID-associated symptoms or the presence of fever as criteria requiring intense telephone or in-personsta monitoring for the following week. Only three patients met criteria. A total of 219/353 had emailsurveys sent. Of 63 patients (28.8%) who returned email surveys by June 10, 2020, 35.9% had alreadymanaged a "pain attack" at home 4 or more times in the prior 12 months, and 45.5% of these saidtheir bad ER experiences were very or somewhat important in that decision. In the prior 14 days, although 30/64 reported a crisis for at least one day, only 4/64 had visited the EmergencyDepartment for pain. On a 0-10 scale, 21/61 patients endorsed "0" for worry that they would beCOVID-infected by going for medical care (weighted mean 3.9), but 18/59 endorsed "10" for worrythey were more at risk of COVID because of SCD (weighted mean 6.31), and 22/60 endorsed "10" forworry they would fare worse than others if COVID infected (weighted mean 6.97). Many patientsforwent "needed" care (16/62) or delayed "needed" care by at least a day (36/61). Eleven patients metcriteria for moderately severe to severe depression on the PHQ-9, and 28/63 somewhat or stronglyagreed with the statement "death is always on the back of my mind" on the SCSS-A. Conclusions In adolescents and adults with SCD, many were already reticent to come to the ED for pain, but asignificant portion reported delays or avoidance of needed care during the early stages of the US COVID pandemic, and few reported using the ED despite over half reporting at least one crisis day in14. Patients nonetheless reported very few COVID-associated symptoms. Fears of COVIDinfection/susceptibility may limit visits for ne ded sickle cell care among adults.

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1. **COVID-19 and SCA: An old friend comes to the rescue**  
   Novelli E. M. Blood 2020;135:1925-1926.

In this issue of Blood, DeBaun1 proposes a forward-thinking evidence-based strategy to limit the impact of the SARS-CoV-2 coronavirus disease (COVID-19) pandemic on children with sickle cell anemia (SCA) at high risk for stroke. DeBaun argues that rapid initiation of low- and fixed-dose (10 mg/kg per day) hydroxyurea treatment while the children are receiving prophylactic chronic transfusions, and before blood supply is disrupted, ensures that when blood shortages do occur, the clinical benefit of hydroxyurea will be more quickly established, and the children will be better protected.Copyright © 2020 American Society of Hematology. All rights reserved.

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1. **COVID-19 and thalassaemia in Iran**  
   Dehshal M. H. Thalassemia Reports 2020;10:25-28.

Coronavirus disease 2019 (COVID-19) has had and continues to have a significant medical, public health, social and economic impact on every society around the world. Some groups of chronic patients including thalassaemia and other haemoglobin disorders were considered from the beginning of the pandemic, as vulnerable and high risk ones with regards to a more severe clinical outcome of the infection with severe acute respiratory syndrome-coronavirus-2 (SARS-CoV-2). This is because patients with thalassaemia can present with many and multiple co-morbidities including diabetes, heart, liver, endocrine and other conditions mainly secondary to iron overload and consequent to ineffective or suboptimal medical care and/or adherence to chelation treatment in particular. Transfusion dependent patients with B-thalassaemia have been greatly affected across the world, including in Iran, a country geographically situated in the so called thalassaemia belt. Iran with about 20,000 patients with B-thalassaemia and quite successful disease specific prevention and management national programmes faced challenges similar to others. Blood shortages for example consequent to COVID-19 precaution measures taken in every country to contain the virus and the difficulties in accessing drugs including lifesaving ones (iron chelation medication) constitute major challenges. In Iran however, and despite the multiple difficulties as described above, SARS-CoV-2 had a rather small impact regarding infection rates as compared to the rest of the countries, albeit a higher mortality rate reaching 26.5% amongst COVID-19 diagnosed patients. More comprehensive data however from a bigger number of patients with thalassaemia across the world infected with SARS-CoV- 2 is necessary to draw any reliable conclusions as to the level of vulnerability to SARS-CoV-2 and importantly the clinical impact of this virus in these patients.Copyright © the Author(s), 2020.

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1. **COVID-19 and thalassaemia: A position statement of the Thalassaemia International Federation**  
   Farmakis Dimitrios European journal of haematology 2020;105:378-386.

OBJECTIVES: Many patients with haemoglobinopathies, including thalassaemia and sickle cell disease, are at increased risk of developing severe complications from the coronavirus disease 2019 (COVID-19). Although epidemiologic evidence concerning the novel coronavirus (SARS-CoV-2) infection in these patients is currently lacking, the COVID-19 pandemic represents a significant challenge for haemoglobinopathy patients, their families and their attending physicians., METHODS: The present statement summarizes the key challenges concerning the management of haemoglobinopathies, with particular focus on patients with either transfusion-dependent or non-transfusion-dependent thalassaemia, identifies the gaps in knowledge and suggests measures and strategies to deal with the pandemic, based on available evidence and expert opinions. Key areas covered include patients' risk level, adaptation of haemoglobinopathy care, safety of blood transfusions, blood supply challenges, and lifestyle and nutritional considerations., CONCLUSIONS: The proposed measures and strategies may be useful as a blueprint for other disorders which require regular hospital visits, as well as for the timely adaptation of patient care during similar future pandemics. Copyright © 2020 John Wiley & Sons A/S. Published by John Wiley & Sons Ltd.

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1. **COVID-19 and the pulmonary complications of sickle cell disease**  
   Sivalingam Thivya EJHaem 2020;1:545-547.

Sickle cell disease (SCD) patients are commonly affected by pulmonary complications such as acute chest syndrome (ACS), pulmonary embolism (PE) and pneumonia that contribute to significant mortality risks. With a greater susceptibility to infection, they are deemed to be vulnerable patients during the current COVID-19 pandemic. In emerging small case studies of SCD patients with COVID-19 and further complicated by pneumonia, ACS, and/or PE, the clinical benefits of early exchange transfusion and Tocilizumab are evident. However, further clinical trials and larger cohort studies are essential to evaluate effective diagnostic and management options for this high-risk group. Copyright © 2020 The Authors. eJHaem published by British Society for Haematology and John Wiley & Sons Ltd.

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1. **COVID-19 in benign hematology: emerging challenges and special considerations for healthcare professionals**  
   Noun Peter Expert review of hematology 2020;13:1081-1092.

INTRODUCTION: Many patients with inherited or acquired benign hematological disorders are at increased risk of developing severe complications from COVID-19. These patients, therefore, require specific advice regarding isolation and changes to their usual treatment schedules. Their disease can also be associated with significant burden, and they necessitate life-long and regular access to therapy, and regular follow-up consultations and hospital visits. The current COVID-19 pandemic is therefore presenting many challenges for these patients, their families, and health-care professionals., AREAS COVERED: This review provides an overview of the reported COVID-19 cases in the literature in patients with certain benign hematological disorders including thalassemia, sickle cell disease, hemophilia, immune thrombocytopenia, venous thromboembolism, and aplastic anemia. The review also outlines some recommendations on how to manage these patients if they are infected with SARS-CoV-2. To review the literature on benign hematological disorders and COVID-19, a bibliographic search was performed using PubMed for articles published between January 2020 and June 2020., EXPERT OPINION: International efforts must be made to continue reporting and better understanding the effects of SARS-CoV-2 infection in these patients and accordingly develop a set of recommendations to optimize the treatment of future infected patients.

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1. **COVID-19 in Patients With Hematologic-Oncologic Risk Factors: Complications in Three Patients**  
   Tafti Dawood Cureus 2020;12:e12064.

The novel COVID-19 infection has demonstrated a spectrum of complications involving vascular, inflammatory, infectious, and metabolic conditions. These complications range from mild loss of smell to more severe acute respiratory distress syndrome (ARDS). Patients with more severe complications often require sedation and mechanical ventilation. Growing research has revealed the role of active malignancy and disease-in-remission status as possible risk factors contributing to the morbidity and mortality in COVID-19 patients. In our descriptive case series, we present three unique cases of complicated COVID-19 infection in patients with hematologic-oncologic risk factors and review the imaging features of their complications. The first patient was a 33-year-old male with sickle cell trait who developed rhabdomyolysis and myonecrosis of the paraspinal muscle in the setting of a physical fitness test; he subsequently developed an abscess at this site, presumably exacerbated by the hypoxemic state of his COVID-19 pneumonia. Our second patient was a 37-year-old male with COVID-19 pneumonia and a history of stage IV Non-Hodgkin's lymphoma in remission who developed spontaneous pneumomediastinum in the absence of positive pressure ventilation. The third COVID-positive patient was a 54-year-old male with a past medical history significant for grade 1 follicular non-Hodgkin's lymphoma in remission with sputum culture positive for mycobacterium avium complex and bronchoscopy positive for candida growth. 18-FDG/PET imaging was performed and demonstrated diffuse intense uptake throughout the lungs reflecting both the COVID-19 pneumonia and the multimicrobial superinfection. Copyright © 2020, Tafti et al.

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1. **Covid-19 in pediatric hematology and oncology patients in New York City**  
   Gampel B. Pediatric Blood and Cancer 2020;67:No page numbers.

Background and Aims: Little is known about Covid-19 disease in pediatric hematology, oncology and hematopoietic cell transplant (HCT) patients. New York City (NYC), the epicenter of Covid-19 infection in the United States, can provide valuable data in this population. We describe the characteristics and clinical course of Covid-19 disease in patients treated at our centers. Method(s): We identified all patients tested for Covid-19, ages 0-21 years, and cared for by pediatric hematology, oncology, and HCT services atMemorial Sloan Kettering Cancer Center (MSK) and New York Presbyterian Hospital (NYP, Columbia University Irving Medical Center and Weill Cornell Medical Center) through April 6, 2020. Demographic, clinical course, and outcomes were identified and descriptively summarized for Covid-19 positive patients. Result(s): Patients (n=174) were tested for Covid-19 at MSK (n=120, all patients were screened) and NYP (n = 54, 'at risk' patients were screened); nineteen tested positive (11%), males > females (adjusted Odds Ratio 3.1). Three had non-malignant hematologic diagnoses, fourteen had cancer, and two were post-HCT. The most common presenting symptoms were fever (n=13), cough (n=9), and difficulty breathing (n=7). Nine of fourteen cancer patients (64%) experienced treatment delays due to Covid-19 positivity. Eleven patients were hospitalized; five required intensive care, including one with sickle cell disease who died of Covid-19 related complications. No Covid-19 positive patients who were initially managed as outpatients subsequently required admission. Conclusion(s): The majority of patients had mild disease and many were managed as outpatients. Males appeared to have a higher incidence of infection compared to females. Covid-19 led to treatment delays in most cancer patients, however those who received myelosuppressive chemotherapy during and prior to infection did not appear to have increased complications. Oncologic patients without underlying comorbidities beyond their cancer do not appear to have a greater risk of complications from Covid-19 than other children.

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1. **COVID-19 in pediatric oncology from French pediatric oncology and hematology centers: High risk of severe forms?**  
   Andre Nicolas Pediatric blood & cancer 2020;67:e28392.

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1. **COVID-19 Infection and Acute Pulmonary Embolism in an Adolescent Female With Sickle Cell Disease**  
   Kasinathan Sushma Cureus 2020;12:e12348.

A previously healthy 20-year-old female presented to the emergency room in April 2020 with complaints of shortness of breath, chest pain, and cough. She was diagnosed with coronavirus disease 2019 (COVID-19) infection and pulmonary embolism (PE). Workup for anemia led to the diagnosis of sickle cell disease (SCD). Patients diagnosed with COVID-19 are at an increased risk for the development of PE and venous thromboembolism (VTE). Anticoagulation prophylaxis and escalation to treatment dosing are recommended in patients admitted with moderate to severe symptoms of COVID-19. PE and VTE are relatively uncommon in the pediatric and adolescent population. Most commonly, patients are diagnosed with thrombophilia or have an underlying hypercoagulable state such as with SCD. Also, symptoms of COVID-19 infection, acute chest syndrome (ACS), and PE can have overlapping features. In this report, we present a case of a late adolescent female with SCD, who was diagnosed with COVID-19, and whose condition was complicated with PE. Copyright © 2020, Kasinathan et al.

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1. **COVID-19 outcomes in individuals with sickle cell disease and sickle cell trait compared to blacks without sickle cell disease or trait**  
   Singh A. Blood 2020;136:54-56.

Introduction: By August 1, 2020 in the United States, more than 3 million cases of Coronavirus disease 2019 (COVID-19) had been reported with more than 150,000 deaths due to this disease. Growing evidence suggests that individuals with the pre-existing conditions of hypertension, diabetes, cardiovascular disease and obesity are at a higher risk of more serious COVID-19 illness. However, the impact of COVID-19 on individuals with sickle cell disease and sickle cell trait as compared to those without sickle cell disease or trait is not known. The objective of this study was to determine the rate of hospitalization, disease symptoms and deaths due to COVID-19, in patients with sickle cell disease and sickle cell trait compared to Blacks without sickle cell disease or trait. Method(s): We leveraged existing electronic health record (EHR) data from multiple sites that contribute data to a research network, TriNetX. TriNetX query platform was used to identify patients with COVID-19 infection based on ICD diagnoses codes or a positive COVID-19 result from a nucleic acid amplification with probe-based detection test, present any time after January 20, 2020 (this is when the first COVID-19 case was detected in the United States) within the patients' EHR data. We report rates of specific COVID-19 related outcomes among individuals with sickle cell disease and trait, calculated as % of patients in cohort with the particular outcome. Our outcomes of interest included COVD-19 related symptoms, hospitalization, and death, which occurred within 2 weeks of COVID diagnosis. We used propensity score matching (greedy nearest-neighbor matching algorithm with a caliper of 0.1 pooled standard deviations) to create balanced cohorts for comparing outcomes between individuals with sickle cell disease or trait and Blacks without sickle cell disease or trait. Risk ratios and risk differences are reported along with 95% confidence intervals. Given multiple outcomes of interest, we considered a more stringent two-sided alpha of less than <0.01, based on a z-test, to determine statistical significance for differences in outcome rates. Result(s): As of July 15, 2020, there were 122 COVID-19 patients who had sickle cell disease and 172 COVID-19 patients who had sickle cell trait. Our comparator groups included 15,762 Blacks who were diagnosed with COVID-19 but did not have sickle cell trait/disease. COVID-19 patients with sickle cell disease were significantly younger and a higher proportion had asthma, type 1 diabetes and preexisting liver conditions compared to Blacks without sickle cell trait/disease (Table 1). COVID-19 patients with sickle cell trait were significantly younger, a higher proportion were females, overweight/obese, and a higher proportion had asthma or type 1 diabetes compared to Blacks without sickle cell trait/disease (Table 1). The rate of respective outcomes for the three groups is shown in Figure 1. Propensity score matching yielded a cohort of patients such that there were no significant differences in demographic and clinical characteristics between patients with sickle cell disease/trait compared to Blacks without sickle cell trait/disease. After matching, COVID patients with sickle cell disease remained at a higher risk of hospitalization, pneumonia and pain compared to Blacks without sickle cell trait/disease (Table 2). The case fatality rates were not significantly different between those with sickle cell disease compared to Blacks. There were no significant differences in COVID outcomes between sickle cell trait and Blacks without sickle cell trait/disease, within the matched cohort. Conclusion(s): These data provide evidence that sickle cell disease imposes additional risk of severe COVID-19 illness and hospitalization, after balancing for age, gender and other preexisting conditions. The death rate between sickle cell disease and Blacks without sickle cell trait/disease was not significantly different. There are no significant differences in COVID-19 outcomes between sickle cell trait and Blacks without sickle cell trait/disease, after balancing for age, gender and other pre-existing conditions.

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1. **Healthcare utilization among children and adolescents with sickle cell disease during the COVID-19 pandemic**  
   Nowlin A. Blood 2020;136:30-31.

Background Early experience with the COVID-19 pandemic showed disproportionately high morbidity and mortality among individuals with certain chronic medical conditions. Individuals with sickle cell disease (SCD) are at high risk for pulmonary and other complications including acute chest syndrome (ACS) and have high rates of hospitalization from other viral respiratory infections, raising concern that COVID-19 would be associated with higher morbidity, mortality and health care utilization among those with SCD. Public health interventions such as social distancing, avoidance of large group activities, and widespread use of masks have been shown to reduce the transmission of COVID-19 in the general population but have been inconsistently implemented. In Georgia, COVID-19 restrictions, including school closures, were implemented in mid-March, and on-site school instruction was replaced by virtual instruction for the remainder of the school year. At our institution, most routine, non-urgent outpatient clinic visits were cancelled or postponed from mid-March through May in order to minimize COVID-19 exposure risk. Efforts to initiate the use of telemedicine as an alternative to in-person office visits were rapidly instituted. We hypothesized that adherence to public health restrictions, especially sheltering in place, would be high among patients and families with SCD, and sought to measure the impact of COVID pandemic on healthcare utilization in children and adolescents with SCD in the Atlanta area. Methods The SCD Program at Children's Healthcare of Atlanta (CHOA) provides comprehensive outpatient, emergency and inpatient services at 3 locations in metropolitan Atlanta. CHOA's Sickle Cell Clinical Database (SCCD) contains prospectively collected demographic, diagnostic, treatment and other clinical information on all patients with SCD beginning in 2010, including all outpatient clinic, emergency department (ED) and inpatient hospital utilization. To assess the impact of COVID-19 on healthcare utilization, we tracked clinic, ED and inpatient utilization for the 4-month period (March through June) 2020 compared with the same 4-month period in 2018 and 2019. Results The figure shows utilization patterns for each four-month period from 2018-2020. As expected, face to face outpatient clinic visits fell dramatically from February to April 2020 (-25% in March, -64% in April) and then returned to pre-COVI D levels by June. The addition of telemedicine visits raised total outpatient visits in June 2020 to above pre-COVID levels. Total utilization during the 4-month period in 2020 were compared to the mean for the same periods in 2018 and 2019. Face to face clinic visits decreased from 2971.5 to 2023 (-32%), ED visits from 1,217 to 687 (-44%), and total inpatient admissions from 699 to 410 (-41%). Admissions with a primary discharge diagnosis of pain decreased from a mean of 407 in 2018-2019 to 173 (-57%), fever/infection from 67.5 to 40 (-41%), and ACS from 101 to 75 (-26%). Patients with chronic pain and/or history of high utilization (>5 admissions in a given year) showed decreases in utilization similar to all other patients. Summary These data describe the significant changes in utilization among pediatric patients with SCD during the COVID-19 pandemic. Face to face outpatient clinic visits decreased during March and April but returned to pre-COVID levels in June. Unexpectedly, ED and inpatient hospital utilization for acute illness decreased dramatically through April and remained low through June. In March there was a significant decrease in the clinic setting due to a large number of cancelled or rescheduled outpatient visits, despite many being rescheduled as telemedicine visits. However, the largest unexpected decrease was seen in emergency department visits and hospitalizations for acute events, specifically fever and pain events. It is also important to note the decreased utilization of patients with chronic pain who are typically high utilizers. During clinic encounters, families mentioned that less stress from s hool, reduced respiratory infections, and better medication adherence with parents at home, were possible contributors to reduced sickle cell symptoms while sheltering in place. These observations will guide the development of a patient survey with the goal of obtaining qualitative data to explain the reasons for decreased utilization during the pandemic. (Figure Presnted).

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1. **Hospitalization and case fatality in individuals with sickle celldisease and COVID-19 infection**  
   Mucalo L. Blood 2020;136:7-8.

Sickle cell disease (SCD) is an inherited hemoglobinopathy that can aect nearly every organsystem. Individuals living with SCD are at high risk of developing serious infections which can furthertrigger disease related complications and attribute additional morbidity and mortality. In light of theevolving pandemic caused by SARS-CoV-2, the causative agent of COVID-19 disease, and thepotential for future infectious disease epidemics, it is important to understand the impact thatCOVID-19 has on hospitalization rates and mortality in this medically vulnerable population. Theobjective of this study was to describe hospitalization and case fatality rates secondary to COVID-19among individuals living with SCD in dierent age groups and compare these to the generalpopulation. The Medical College of Wisconsin established the international SECURE-SCD Registry to collectdata on pediatric and adult COVID-19 infections in individuals living with SCD. Providers areinstructed to report confirmed COVID-19 cases to the registry after suicient time has passed to observe the disease course through resolution of acute illness and/or death. For each case, providerscomplete a short form that includes the following data: patient demographics, COVID-19 relatedhospitalization, COVID-19 severity/management strategies, if the patient died due to COVID, andother information about SCD complications. Data are de-identified and without protected healthinformation to facilitate rapid and increased reporting. We calculated the hospitalization rate andcase fatality rate for individuals with SCD by specific age group and contrasted it with the ratespublicly available for the general Black population. We utilized data from California Department ofPublic Health for case fatality rate comparison in Blacks and data from COVID-NET forhospitalization rate comparison. We used indirect age adjustment to calculate standardized mortalityratios using COVID-19 data from California state as the reference population. As of July 17th 2020, 218 cases of COVID-19 in Blacks with SCD in the US were reported to theregistry. There was a slight predominance of females (52.8%) and 32.1% of reported cases werepatients 18 years and under. There were 15 deaths reported with overall mortality rate of 6.9%. Figure1 shows the distribution of cases and deaths by age group and gender. Mortality rate in SCD patientswas highest in the 50-64 years age group (23.1%) in contrast to mortality rate peaks seen in thegeneral population in patients older than 80 years (Table 1). Young adult SCD patients aged 18-34years had a case fatality rate of 3.3% and those aged 34-50 years had a rate of 14.9%. CaliforniaDepartment of Public Health report case fatality rates for Blacks are less than 1% in both of thesecomparative age groups. Age-standardized mortality ratio shows that individuals with SCD are 7.7times more likely to die due to COVID-19 infection compared to the general population. The overallhospitalization rate in individuals with SCD was 72.5% and 18.8% of reported hospitalized cases werechildren. Among hospitalized adults with SCD, stratification by age showed that 85% were aged 18-49, whereas only 25.7% of people 18-49 years in the general Black population were hospitalized(Table 2). Our findings show that individuals with SCD who have COVID-19 infection have higher rates of deathdue to COVID-19 than the general Black population. Also, a large proportion of COVID hospitalizationfor the SCD population occurs among the younger age group. Further analysis is planned to examineeects of underlying comorbidities and prior SCD-associated complications on the severity ofCOVID-19 in individuals with SCD.

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1. **How to treat and manage covid19 in SCD patients**  
   Verdiyeva N. Hematology, Transfusion and Cell Therapy 2020;42:76.

Objective: Coronavirus disease 2019 (COVID-19) is an infectious disease caused by severe acute respiratory syndrome coronavirus 2 (SARS-CoV-2). It was first identified in December 2019 in Wuhan, China, and has resulted in an ongoing pandemic. Case report: A 24-year-old man with a history of SCD (HbS/beta0-thalassemia) on maintenance hydroxyurea therapy presented to our hospital, with a complaint of pain in the extremities and chest over two days. The patient with mild cough and high fever was hospitalized. Blood tests and lung CT were performed. Result of blood test show evidence of systemic hemolysis with a decrease in hemoglobin from 8.9 g/dL to 6.7 g/dL. His white blood cell count was 25.2 x 103/muL, CRP 243.21 mg/L. CT scans of the lungs showed a consolidated area where air bronchograms were observed in and around the medial segment of the middle part of the right lung and the posteriobasal segment of the lower part of both lungs, and an icy glass landscape was observed. Lung damage is 1-5% (grade I). His oxygen saturation SpO2 was normal (98%). The SARS-CoV-2 PCR nasopharyngeal swab testing was sent and returned negative on hospital day one after which the patient was started on antiviral and antibiotic for severe COVID-19 pneumonia. An improvement in blood counts was observed 4 days after starting treatment (WBC 16.93 x 103/muL, CRP 100.31 mg/L). On day ten, after normalization of all symptoms and blood values the patient was discharged home. Methodology: In this study we selected 1 patient with SCD followed in Thalassemia Center of Azerbaijan. Result(s): Given the higher likelihood of ACS it is possible that SCD patients are also at higher risk of such complications from COVID-19, particularly those with a history of pulmonary comorbidities. However, it is unclear if the SARS-CoV-2 pandemic will lead to increased rates of ACS for sickle cell patients. Still, hospitalized sickle cell patients should be monitored closely for development of ACS and if this occurs, exchange transfusion should be promptly initiated. Conclusion(s): COVID-19 pneumonia as a cause of acute chest syndrome in an adult sickle cell patient. Patients with sickle cell disease (SCD) who are infected with COVID-19 may have a significant risk of developing acute chest syndrome (ACS), a potentially life-threatening complication. In this case we will present how manage COVID 19 in patient with SCD.Copyright © 2020

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1. **Impact of a national lockdown for COVID-19 on morbidity andmortality among children with sickle cell anaemia at a tertiarycare hospital in Uganda**  
   Namazzi R. Blood 2020;136:33-34.

COVID-19 and its prevention has put considerable strain on health care systems in low and middle-income countries (LMIC). In Uganda, a national lockdown was declared on March 18, 2020, inresponse to COVID-19 pandemic and concern of spread of cases without aggressive measures toprevent spread. The lockdown consisted of closure of all offices except essential ones, orders to stayat home unless an emergency occurred, school closure, a ban on all meetings of more than 10people, a ban on public and private transport, closing down of all shops, malls, restaurants, places ofworship and other facilities in which group meetings might occur, keeping a distance of at least 2metres from other people in public places and a 7:00 p.m. to 6:30 a.m. curfew. Hospitals howeverremained open and operational. We describe the impact of the lockdown in Uganda in response tothe COVID-19 pandemic on the morbidity and mortality in children with sickle cell anaemia (SCA) ata tertiary hospital in Uganda. The number of clinic visits for SCA related complications and deathwere compared in the pre-lockdown (November 2019 to February 2020) and during COVID-19lockdown periods (March 2020 to June 2020) in children aged 1- 4.99 years enrolled in a SCAresearch study [Zinc for Infection Prevention in Sickle cell anaemia (NCT03528434)] at Jinja Hospital,Uganda. In the study, children with SCA are asked to return to the hospital for evaluation wheneverthey are unwell. Follow up phone calls are made to ascertain the wellbeing of the children and identify any who are unable to come to the hospital. During the lockdown, follow up calls continuedand facilitation was provided for caregivers to bring any child who was unwell to the hospital forevaluation. A total of 238 children with a mean (standard deviation) age of 2.7(1.1) years were enrolledand were being followed up when the pandemic started. The incidence of hospital sick visits pre-lockdown and during the lockdown period was 7.7 vs 4.0 person-year, (p= <0.0001). Incidence ofhospitalization, pain crises, severe anaemia, or malaria were all higher in the pre-lockdown periodthan during the lockdown period, 2.4 vs.1.0, 1.8 vs. 0.7, 0.7 vs. 0.4, 0.6 vs. 0.2 and per person yearrespectively (all p values < 0.01). There were no deaths during the lockdown period compared to 1death in the pre-lockdown period. Less than 1000 cases of COVID-19 were reported nationally in thisperiod, and none of the study children had known COVID-19 infection, though testing capacity forthis was limited. In this cohort of children with SCA, hospitalization and morbidity from SCA-relatedcomplications and malaria were are significantly lower during a lockdown period for COVID-19pandemic than before the lockdown. Reduced access to hospital care is unlikely to explain thesefindings, as sick children still received care at the hospital, and there was no increase in mortality.Reduced interaction with peers because of the lockdown and social distancing, leading to fewerinfections that may trigger SCA-complications, may explain the reduced incidence of SCAcomplications in this population during the COVID-19 lockdown period in Uganda.

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1. **Impact of sickle cell trait on morbidity and mortality from SARS-COV-2 infection**  
   Merz L. E. Blood 2020;136:31-32.

Introduction: The high morbidity and mortality of SARS-CoV-2 in Blacks or African American in the United States is well established. Individuals with sickle cell trait (SCT), who are mostly Black or African American, have adverse health outcomes in situations of increased physiologic stress. The novel corona virus SARS-CoV-2 causes a severe multi-systemic viral infection that induces intense inflammation and metabolic derangements that can exacerbate RBC sickling and organ damage. The purpose of this study is to evaluate the impact of SCT status on the outcome of patients hospitalized for SARS-CoV-2. Method(s): We conducted a multi-center, IRB-approved, retrospective analysis of Black/African American patients who were admitted for management of SARS-CoV-2 infection from March 24, 2020 to June 2, 2020. Patients were identified using an electronic medical record (EMR) report that selected for race as "Black or African American" and a positive SARS-CoV-2 PCR test during that admission. We excluded patients admitted for reasons other than SARS-CoV-2 infection and reviewed only the in-hospital experience. Patient demographics, co-morbidities, admission laboratory values, complications of SARS-CoV-2 infection, and status on discharge were abstracted by manual chart review. High performance liquid chromatography (HPLC) was performed on discarded blood of patients to test for sickle cell trait. The primary objective was to evaluate the impact of SCT status on morbidity outcomes of Blacks/African Americans hospitalized with SARSCoV- 2 infection. Categorical data were tested using the Fisher exact test, and quantitative data were tested using the Wilcoxon rank sum test. Testing was done at the nominal 0.05 two-sided significance level. Result(s): One hundred and sixty-six Black or African American patients admitted for SARS-CoV-2 infection are included in the analysis. Twenty patients had SCT, 143 had normal hemoglobin (AA) and 3 had hemoglobin C trait (AC). The 146 patients with AA and AC hemoglobin were pooled together. Patient demographics, comorbidities, and lab values on admission by SCT status are shown in Table 1. Complications of SARS-CoV-2 by SCT status is shown in Table 2. Among Black or African American patients admitted for SARS-CoV-2 in this study, SCT represented 12.0% of the total. At the time of admission, individuals with SCT had significantly higher creatinine (p=0.004) but were less likely to present with a history of chronic lung disease (p=0.004). However, a history of chronic lung disease was not in itself associated with death in hospital, p=0.056, and creatinine at admission was not associated with death, p=0.483. Correspondingly, a total of 19 of the 146 patients without SCT died in the hospital (13%), compared to 3 of 20 SCT patients (15%), Fisher exact test p-value = 0.732, despite the difference in the groups at admission. Discussion(s): The higher morbidity and mortality from SARS-CoV-2 infection in Black or African Americans is well-documented. This study showed equivalent outcomes in patients admitted for SARS-CoV-2 infection whether, or not they had SCT. There was a lower rate of chronic lung disease in patients with SCT, but no difference in respiratory outcome from SARS-CoV-2 between the groups. Patients with SCT also had worse creatinine at presentation but there was no difference in hospital death or end organ complications at discharge. Notably, individuals with SCT made up 12% of Black patients admitted for SARS-CoV-2 in this study which is higher than the reported prevalence of SCT of 7.31% in the African American population (p=0.025), but this is of unclear significance. Our study is limited by the restriction to one metropolitan area and by being retrospective in nature, but the initial data suggests that those with SCT may be more frequently admitted to hospital when infected by SARS-CoV-2 than individuals without SCT. A larger prospective study across multiple regions of the United States should be considered to further assess the prevalence of SARS-CoV-2 in the African American co munity and the apparent increased rate of hospitalizations for SARS-CoV-2 infection in individuals with SCT. (Table Presented).

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1. **Implications of COVID-19 infections in sickle cell disease**  
   John Nitin Ashok The Pan African medical journal 2020;36:81.

Sickle cell disease is a major concern of public health significance in Africa. Nearly 2/3rd of the global burden of sickle cell disease (SCD) is found to be in sub-Saharan Africa. There is increased mortality risk in sickle cell disease patients in Africa due to associated complications such as acute chest syndrome, asthma, pulmonary emboli and sepsis. Sickle cell disease management is the major contributor of financial burden on the government. Moreover, there is a shortage of medical specialists in Africa. COVID-19 pandemic has further led to devastating impact on economy and health globally. The chances of SCD patient contracting COVID-19 infections are higher as these patients are immunocompromised and may be at a higher risk of mortality. Practicing preventive measures including isolation and social distancing by these patients will prevent mortality rates as well as economic burden on government in the present unprecedented COVID-19 pandemic. Copyright © Nitin Ashok John et al.

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1. **Initiating adjunct low-dose hydroxyurea therapy for stroke prevention in children with SCA during the COVID-19 pandemic**  
   DeBaun Michael R. Blood 2020;135:1997-1999.

[Available online at this link](https://www.knowledgeshare.nhs.uk/index.php?PageID=link_resolver&link=3bb0044f46626eb30a0a54e99017668c)

1. **Mitigating the effect of the COVID-19 pandemic on sickle cell disease services in African countries**  
   Dexter Daniel The Lancet. Haematology 2020;7:e430-e432.

[Available online at this link](https://www.knowledgeshare.nhs.uk/index.php?PageID=link_resolver&link=f79ad528de5f48e265625b595d07d720)

1. **Preliminary Data on COVID-19 in Patients with Hemoglobinopathies: A Multicentre ICET-A Study**  
   de Sanctis Vincenzo Mediterranean journal of hematology and infectious diseases 2020;12:e2020046.

Objectives: This study aims to investigate, retrospectively, the epidemiological and clinical characteristics, laboratory results, radiologic findings, and outcomes of COVID-19 in patients with transfusion-dependent beta thalassemia major (TM), beta-thalassemia intermedia (TI) and sickle cell disease (SCD)., Design: A total of 17 Centers, from 10 countries, following 9,499 patients with hemoglobinopathies, participated in the survey., Main outcome data: Clinical, laboratory, and radiologic findings and outcomes of patients with COVID-19 were collected from medical records and summarized., Results: A total of 13 patients, 7 with TM, 3 with TI, and 3 with SCD, with confirmed COVID-19, were identified in 6 Centers from different countries. The overall mean age of patients was 33.7+/-12.3 years (range:13-66); 9/13 (69.2%) patients were females. Six patients had pneumonia, and 4 needed oxygen therapy. Increased C-reactive protein (6/10), high serum lactate dehydrogenase (LDH; 6/10), and erythrocyte sedimentation rate (ESR; 6/10) were the most common laboratory findings. 6/10 patients had an exacerbation of anemia (2 with SCD). In the majority of patients, the course of COVID-19 was moderate (6/10) and severe in 3/10 patients. A 30-year-old female with TM, developed a critical SARS-CoV-2 infection, followed by death in an Intensive Care Unit. In one Center (Oman), the majority of suspected cases were observed in patients with SCD between the age of 21 and 40 years. A rapid clinical improvement of tachypnea/dyspnea and oxygen saturation was observed, after red blood cell exchange transfusion, in a young girl with SCD and worsening of anemia (Hb level from 9.2 g/dl to 6.1g/dl)., Conclusions: The data presented in this survey permit an early assessment of the clinical characteristics of COVID 19 in different countries. 70% of symptomatic patients with COVID- 19 required hospitalization. The presence of associated co-morbidities can aggravate the severity of COVID- 19, leading to a poorer prognosis irrespective of age.

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1. **Protecting vulnerable patients with inherited anaemias from unnecessary death during the COVID-19 pandemic**  
   Roy Noemi B. A. British journal of haematology 2020;189:635-639.

With the developing COVID-19 pandemic, patients with inherited anaemias require specific advice regarding isolation and changes to usual treatment schedules. The National Haemoglobinopathy Panel (NHP) has issued guidance on the care of patients with sickle cell disease, thalassaemia, Diamond Blackfan anaemia (DBA), congenital dyserythropoietic anaemia (CDA), sideroblastic anaemia, pyruvate kinase deficiency and other red cell enzyme and membrane disorders. Cascading of accurate information for clinicians and patients is paramount to preventing adverse outcomes, such as patients who are at increased risk of fulminant bacterial infection due to their condition or its treatment erroneously self-isolating if their fever is mistakenly attributed to a viral cause, delaying potentially life-saving antibiotic therapy. Outpatient visits should be minimised for most patients, however some, such as first transcranial dopplers for children with sickle cell anaemia should not be delayed as known risk of stroke will outweigh the unknown risk from COVID-19 infection. Blood transfusion programmes should be continued, but specific changes to usual clinical pathways can be instituted to reduce risk of patient exposure to COVID-19, as well as contingency planning for possible reductions in blood available for transfusions. Bone marrow transplants for these disorders should be postponed until further notice. With the current lack of evidence on the risk and complications of COVID-19 infection in these patients, national data collection is ongoing to record outcomes and eventually to identify predictors of disease severity, particularly important if further waves of infection travel through the population. Copyright © 2020 British Society for Haematology and John Wiley & Sons Ltd.

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1. **Rare Anaemias, Sickle-Cell Disease and COVID-19**  
   Vives Corrons Joan-Lluis Acta bio-medica : Atenei Parmensis 2020;91:216-217.

For rare haematological diseases (RHD), the first question to be answered is if patients with be- nign red blood cell (RBC) defects like haemoglobinopathies, membranopathies and enzymopathies are more vulnerable to COVID-19 infection. Up to now, there is no yet literature on the subject, but, like in general population, the presence of comorbidities such as diabetes, heart disease, pulmonary hypertension, reduced kidney and/or liver function, worsen the effects of the infection. Splenectomy may be an additional risk factor.

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1. **Red blood cell exchange to avoid intubating a COVID-19 positive patient with sickle cell disease?**  
   Allison David Journal of clinical apheresis 2020;35:378-381.

As the COVID-19 pandemic continues to claim lives across the globe, insufficient data exists regarding the optimal treatment. It is well known that patients 55 years of age or older and patients with certain chronic diseases are at higher risk of severe illness, including acute respiratory distress syndrome and death. A potentially fatal pulmonary complication of sickle cell disease, acute chest syndrome, can be precipitated by acute infections, including respiratory viruses. We report the case of a patient with sickle cell disease (HbSC) who developed COVID-19 pneumonia and acute chest syndrome who was treated with emergent red blood cell exchange in order to avoid endotracheal intubation. Copyright © 2020 Wiley Periodicals LLC.

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1. **Remodelling of specialist services enables safe reduction in hospital admissions of patients with sickle cell disease: Lessons from the COVID-19 pandemic**  
   Tsitsikas Dimitris A. Clinical medicine (London, England) 2020;20:e241-e243.

Sickle cell disease is characterised by recurrent painful crises often leading to hospitalisation. During the COVID-19 pandemic, it was important to try to reduce the need for hospital admission for these high-risk patients while at the same time ensuring that hospital avoidance did not put them at risk of deterioration from disease-related complications. In the 3-month period between March and May 2020, there was a significant reduction in the number of hospital admissions as well as mean length of stay compared with the mean figures over the same months in the preceding 5 years (2015-19), with an overall reduction in inpatient days of 77%. There were no cases of unsafe hospital avoidance or presentations to hospital that were inappropriately delayed. Frequent telephone communication with patients and provision of ambulatory care were, among others, two very important means of supporting our patient population. Copyright © 2020 Royal College of Physicians 2020. All rights reserved.

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1. **Severe and fatal forms of COVID-19 in children**  
   Oualha M. Archives de pediatrie : organe officiel de la Societe francaise de pediatrie 2020;27:235-238.

OBJECTIVES: The aim of this study was to describe severe forms of novel coronavirus disease 2019 in children, including patient characteristics, clinical, laboratory, and imaging findings, as well as the disease management and outcomes., METHODS: This was a retrospective, single-center, observational study conducted in a pediatric intensive and high-dependency care unit (PICU, HDU) in an urban hospital in Paris. All patients, aged from 1 month to 18 years, admitted for confirmed or highly suspected SARS-CoV-2 were included., RESULTS: We analyzed the data of 27 children. Comorbidities (n=19, 70%) were mainly neurological (n=7), respiratory, (n=4), or sickle cell disease (n=4). SARS-CoV-2 PCR results were positive in 24 children (nasopharyngeal swabs). The three remaining children had a chest CT scan consistent with COVID-19. Respiratory involvement was observed in 24 patients (89%). Supportive treatments were invasive mechanical ventilation (n=9), catecholamine (n=4), erythropheresis (n=4), renal replacement therapy (n=1), and extracorporeal membrane oxygenation (n=1). Five children died, of whom three were without past medical history., CONCLUSION: This study highlighted the large spectrum of clinical presentation and time course of disease progression as well as the non-negligible occurrence of pediatric life-threatening and fatal cases of COVID-19 mostly in patients with comorbidities. Additional laboratory investigations are needed to further analyze the mechanism underlying the variability of SARS-Cov-2 pathogenicity in children. Copyright © 2020 French Society of Pediatrics. Published by Elsevier Masson SAS. All rights reserved.

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1. **Sickle Cell Disease (SCD) and COVID-19 – A Case Series**  
   Al-Naami Awaji 2020;:No page numbers.

The ongoing pandemic of COVID-19 that started in the Hubei province of China in late December 2019, caused by severe acute respiratory syndrome corona virus-2 (SARS CoV-2). Globally millions affected by the disease so far. The risk of COVID-19 severity and its complications increases with age and other comorbidities. The course of SARS-CoV-2 infection or its related complications has yet to be established in patients with sickle cell disease (SCD), once more evidence is available. It is clear from the available data that the course of COVID-19 in patients with SCD is mild to moderate, seldom severe, and rarely fatal. Herein we report three known cases of SCD with confirmed COVID-19, in whom the course of the disease was mild to moderate and uncomplicated with uneventful recoveries.

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1. **Sickle cell disease related outcomes in patients evaluated forCOVID-19 infections in South Carolina**  
   Noisette L. Blood 2020;136:38-39.

Introduction: The severe acute respiratory syndrome coronavirus 2 (SARS-Cov-2), first identified in Wuhan, China,was declared a pandemic by WHO in March 2020 due to its high transmission rate. Due to the diffusevasculo-endothelial damage, individuals with Sickle Cell Disease (SCD) are at risk to develop severeclinical complications, if infected with coronavirus 19 (COVID-19).[1] Given this risk, a systematicevaluation of individuals with SCD presenting with COVID19 infection is paramount to identify thevariable clinical manifestations and complications encountered in children and adults with SCD. Method(s): A retrospective chart review was conducted from January to June 2020 at the Medical University of South Carolina. We included individuals with sickle cell disease of all genotypes, from 0 to 65 years of age found to be positive for COVID19 by polymerase chain reaction (PCR). Patients' past medicalhistory, clinical presentations, admissions, treatment, complications, and mortality data werereviewed. The data was collected with REDCap @ and descriptive data analysis was conducted perSPSS @ . Results : Of the identified 23 patients with SCD who tested positive for COVID during the time specified, 19(82.6%) had Hgb SS genotype, two had Hgb SC (9%) and two Hgb Sbeta+ thalassemia (9%) with similarincidence in both genders (47.8% male and 52.2 % female). All patients were African American. Themean age was 26.13+/-11.53 years. In the last three years they had admissions for pain at a mean of4.29 +/- 5 and admissions for acute chest syndrome 1+/- 2.2. Six participants (26.1%) had history of mild asthma. Two (8.7%) had pulmonary hypertension. No participants had a history of silent stroke.One participant had history of ischemic stroke, three (13%) had history of pulmonary embolism, andsix (26.1%) had deep vein thrombosis (DVT). A variable clinical presentation was noted in ourpopulation (Table 1). Of the 23, only nine (39%) required admission of which only one met criteria for intensive care (4.3%)requiring respiratory support with high flow nasal canula. All participants recovered well with themean length of admission 4.36+/- 3.8 days. Treatment included supportive care including transfusionsupport, two (8.7%) needed simple transfusion, two (8.7%) needed exchange transfusion. Regardingthe laboratory values, coagulations studies were noted to be elevated among all those obtained, butoverall limited values were obtained. (Table 2) Thus far no complications of stroke, thrombosis, orpulmonary emboli are noted in the patients positive for COVID in sickle cell disease at our institution.No deaths were reported. Conclusion(s): Our population reflects what has been described thus far in other cohorts regarding patientdemographics, clinical presentation and evolution of disease. Missing laboratory results is most likelydue to the mild severity which did not require further clinical evaluation. The absence of VTE/PE maybe explained by the low rate of ICU admissions. A similar ICU admission rate of 13% in the same agegroup as our population was described in a study conducted in France with 83 patients. [2]Compared to a study conducted in Detroit, Michigan, our population underwent comparable rates of transfusions with 3 patients compared to 4 in our population, again most likely due to the mildseverity. [3] Our results reflect only MUSC's testing sites and we are dependent on patient's self-report whichmay not represent our entire population. To address this issue, as part of the Sickle Cell SouthCarolina network, we are partnering with two other institutions to assess SARS-Cov-2 infection inSouth Carolina. SARS-CoV-2 pandemic has brought to light many disparities encountered in the American healthcare system. It is premature to evaluate the immediate and long-term ramifications of COVID19 in individuals with sickle cell disease, due to which we plan to continue to monitor for the next 2 years.

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1. **Sickle cell disease: High risk or no risk for coronavirus disease 2019 infection**  
   Albagshi M. H. Journal of Applied Hematology 2020;11:89-90.

[Available online at this link](https://www.knowledgeshare.nhs.uk/index.php?PageID=link_resolver&link=8b1aa27a8300816948c3bc8b202d62c2)

1. **Sickle cell individuals are less vulnerable for corona virus disease 2019-an enigma**  
   Sonone A. International Journal of Research in Pharmaceutical Sciences 2020;11:1015-1017.

The "Coronavirus Disease 2019 (COVID-19)" caused by "Severe Acute Respiratory Syndrome Coronavirus 2 (SARS-CoV-2)" has emerged in December 2019 and was announced as a pandemic by "World Health Organization". As of today, no specific therapeutics drugs are available for COVID-19. Thus patients have to rely on symptomatic adjuvant therapies. Iron is critical in various physiological processes like DNA/RNA synthesis and generation of ATP. In pathological condition, iron is vital for the host as well as the pathogen Lit-erature search revealed iron chelation therapy is one of the promising and emerging treatment modality for COVID-19. In pathological condition, iron is essential for the host as well as the pathogen like viruses which require intra-cellular iron for replication and propagation. Sickle cell anaemia is hemolytic anaemia, where "Sickle haemoglobin (HbS)" is a structural variant of normal adult haemoglobin. Haemoglobin composed of heam and globulin molecules. An iron atom of heam helps in binding of oxygen molecules. In sickle cell indi-viduals, haemoglobin concentration is reduced. Thus viruses do not succeed in replication and proliferation due to deprived iron concentration; they may be less vulnerable for COVID-19 due to reduced iron load. Another fact is that sickle cell individuals are immune to Malaria due to HbS. Some reports say that the incidence of COVID-19 is less in Malaria population counties. Thus it may be postulated that sickle cell individuals may also develop immunity to SARS-CoV-2, as evidenced by less incidence of COVID-19 in Malaria patients.Copyright © 2020 International Journal of Research in Pharmaceutical Sciences. All rights reserved.

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1. **Sickle cell trait and the potential risk of severe coronavirus disease 2019-A mini-review**  
   Kehinde Tawakalitu Abosede European journal of haematology 2020;105:519-523.

Coronavirus Disease 2019 (COVID-19) pandemic is a rapidly evolving public health problem. The severity of COVID-19 cases reported hitherto has varied greatly from asymptomatic to severe pneumonia and thromboembolism with subsequent mortality. An improved understanding of risk factors for adverse clinical outcomes may shed some light on novel personalized approaches to optimize clinical care in vulnerable populations. Emerging trends in the United States suggest possibly higher mortality rates of COVID-19 among African Americans, although detailed epidemiological study data is pending. Sickle cell disease (SCD) disproportionately affects Black/African Americans in the United States as well as forebearers from sub-Saharan Africa, the Western Hemisphere (South America, the Caribbean, and Central America), and some Mediterranean countries. The carrier frequency for SCD is high among African Americans. This article underscores the putative risks that may be associated with COVID-19 pneumonia in sickle cell trait as well as potential opportunities for individualized medical care in the burgeoning era of personalized medicine. Copyright © 2020 John Wiley & Sons A/S. Published by John Wiley & Sons Ltd.

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1. **Staff risks stratification in preparation for COVID-19 in a tertiary healthcare facility in Nigeria**  
   Obaseki Darlington Ewaen The Pan African medical journal 2020;35:124.

Introduction: this report is a documentation of a staff risk stratification programme, undertaken in University of Benin Teaching Hospital, with outcomes, and the actions taken to protect staff., Methods: an adapted risk stratification tool was circulated to all staff through their respective heads of departments/units. Staff were expected to voluntary assess their health and risk status in the context of COVID-19, using the tool. A central multi-disciplinary screening committee assessed submissions and invited staff who required further evaluation for physical interviews. Respondents were categorized into three risk/exposure groups from lowest to highest - A, B, and C, based on their individual health assessments, occupational exposures, and information obtained from direct interviews., Results: the committee received submissions from 746 staff, representing 19.4% (about a fifth) of the hospital's 3,840 staff. One hundred and twenty two of these were invited for physical interviews, of whom 88 (72.1%) were categorized as high risk (Category C): pregnancy (53.4%); bronchial asthma (19.3%); hypertension (11.4%); cancer (3.4%) and sickle cell disease (2.3%); fractures and pulmonary tuberculosis (1%, respectively). These staff were recommended for redeployment from areas of high risk exposure to COVID-19., Conclusion: a management-driven risk assessment of hospital staff in preparation for the COVID-19 pandemic revealed that a fifth of staff assessed themselves as being vulnerable to adverse outcomes from exposure. It is our hope that similar risk stratification programmes will become standard practice in healthcare facilities during disease outbreaks, especially in Africa. Copyright ©Darlington Ewaen Obaseki et al.

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1. **Thalassaemia prior and consequent to COVID-19 pandemic. The perspective of thalassaemia international federation (TIF)**  
   Eleftheriou A. Thalassemia Reports 2020;10:1-5.

Patients with haemoglobin disorders, particularly beta-thalassaemia or sickle cell disease (SCD) or combined forms, on account of their underlying disease pathology and associated (iron load mainly in the case of thalassaemia) co-morbidities are defined as high-risk individuals prone to develop more severe complications from coronavirus disease-2019 (COVID-19). Despite the fact that epidemiological evidence concerning severe acute respiratory syndrome-coronavirus-2 (SARS-CoV-2) infection in these patients is currently limited across the world, it is expected that COVID-19 pandemic will have a very serious, negative impact on national economies, healthcare and social systems and consequently significant respective repercussions on the patients particularly chronic ones, and their families. Although this may be a temporary challenge in some countries of high HDI and robust health, public health and social infrastructures, this can be a long term challenge with serious to tragic consequences in countries particularly devoid of universally covered heath care systems. Thalassaemia International Federation (TIF) in this present paper summarises the key challenges as expressed byNonthe patients, their families and involved health care professionals themselves prior and consequent to COVID-19 pandemic, describes its response during the pandemic and expresses its position in support of its global patient community.Copyright © the Author(s), 2020 Licensee PAGEPress.

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1. **The impact of COVID-19 on the GOSH newborn screening service**  
   Ramgoolam T. Archives of Disease in Childhood 2020;105:A35.

The outbreak of COVID-19 stalled most clinical and non-clinical services across the country, however the Newborn bloodspot screening (NBS) was rated a 'critical service' because the early detection and treatment of 'at-risk' babies helps to reduce mortality and morbidity. NBS is a national public health programme where babies are screened in the first week of life for 9 conditions; 6 Inherited Metabolic Diseases (IMDs), Congenital Hypothyroidism, Cystic Fibrosis and Sickle Cell Disease. The NBS laboratory at GOSH is the largest in the UK, screening approximately 125,000 babies yearly. Since the start of the pandemic, the NBS service has witnessed many challenges but the team always responded with remarkable resilience and flexibility. Our staffing level was reduced by 21% due to shielding regulations imposed during lockdown. Apart from donning masks and other forms of PPE, COVID-19 forced the laboratory to embrace safer and more sustainable ways of service delivery while ensuring the wellbeing of its staff. We piloted remote working across all laboratory processes and conducted risk assessments to mitigate against any impending risks while actualising established failsafe. As a pioneer for R&D, GOSH NBS laboratory was fully engaged in activities to support the SCID pilot (a rare condition of the immune system) before the first COVID-19 wave struck. The project discussions continued virtually and our laboratory is now set to commence the SCID pilot following the initial postponement. The GOSH Newborn screening laboratory has delivered an uninterrupted service with no delays in the reporting of positive results including reviewing of IMD results at weekends. Our laboratory processes have been adapted and some members of our team have completed training as Peer Support Workers to enable participation in early wellbeing discussions among their colleagues. This has provided us with much needed resilience in the likely emergence of a second-wave.

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1. **Validation of a predictive score of acute chest syndrome (presev-2 study) in adults**  
   Kassasseya C. Blood 2020;136:23.

Introduction Vaso-Occlusive Crisis (VOC), the most common manifestation of sickle cell disease (SCD), is the first cause of death when complicated by an acute chest syndrome (ACS). We previously developed a predictive score for the occurrence of ACS in SCD patients admitted at hospital for a VOC episode (Bartolucci et al., 2016). Two hundred and fifty patients with severe VOC requiring hospitalization were included in the PRESEV I study and 19% developed a secondary ACS within days postadmission (median [IQR] : 3 [2.3]). A multivariate analysis of these data established a predictive score of secondary ACS, with a negative predictive value of 98,9% for the low-level risk. Variables for calculation of the score were: reticulocytes and leucocytes counts, hemoglobin and categorical pain score (spine -pelvis). Interestingly enough, Hydroxyurea treatment did not have any impact on the score. Our goal was to validate this score in a multicenter international study, as it could represent a useful tool for physicians, for improving VOC management, but also be of use for therapeutic trials. Herein, we present results for adults. Methods This international, multicenter, prospective, observational study was performed in thirteen centres, over two continents (Africa and Europe) and five countries (Mali, Togo, England, Belgium and France). Homozygous SCD patients, both adults and children (>2 years), were included, with severe VOC requiring admission at the emergency unit. Severe VOC was defined as pain or tenderness affecting at least one part of the body (e.g. limbs, ribs, sternum, head, spine and/or pelvis) requiring opioids (level 3) and not attributable to other causes. The primary outcome measure was the occurrence of an ACS, defined as an auscultatory abnormality (crepitation or bronchial breathing), or the association of a new radiologic infiltrate and chest pain or decreased breath sounds. Secondary outcome measures were hospital length of stay, morphine consumption, transfusion, hospitalization in intensive care unit and mortality. The following parameters were recorded: temperature, blood pressure, oxygen saturation, respiratory frequency, categorical pain score. Pulmonary auscultation was performed at least once a day by a physician, every day, for the length of stay. The auscultatory abnormality, defined as crepitation, bronchial breathing or decreased breath sounds, was confirmed by a second physician. If a patient was discharged before day 5, follow-up with a phone call and/or a visit, 3 to 7 days after discharge were performed, to prevent complications or ACS. Results Three hundred and seventy-two adult patients with a severe VOC requiring hospitalization were included. Mean age was 29 (+/-8) years old, sex ratio (female/male) was 1.2. A hundred and ninety-one patients came from Europe and 181 from Africa. Out of the 372 patients, 68 (18.3%) further developed a secondary ACS. Mean day of ACS episode was 3.3 (+/-1.37). Among the 304 patients who did not develop an ACS episode (81.7%), 41 had a low risk score of developing a secondary ACS (predictive score 5). Among the 68 patients who developed a secondary ACS, 43 had a high-risk score 11). Results are shown in table 1. Three deaths were reported, all on African continent (ACS, anemia and end-stage nephropathy). Discussion and Conclusion Our multicenter international study has allowed confirmation of the incidence of patients developing a secondary ACS during a VOC. This study confirms that the PRESEV score could indeed represent a S useful tool for physicians, most especially within emergency structures, by identifying patients with a low risk of further developing an ACS with a good NPV and thus, allowing a better management of SCD patients experiencing a VOC. The PRESEV score was successfully put to practice during the Covid-19 pandemic at the referral SCD center of Henri Mondor hospital and identified "low-risk" patients, who benefited from homecare services (DREPADOM). As such, the PRESEV score could represent a significant help when one considers the shortne s of bed availabilities in certain circumstances (pandemics, hospital settings in resource-limited countries, etc.). Finally, the identification of "high-risk" patients could also become useful for improving the feasibility of clinical trials for the prevention of ACS. (Table Presented).

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1. **Venous thromboembolism prophylaxis practices for patients with sickle cell disease pre and during the COVID-19 pandemic**  
   Davila J. G. Blood 2020;136:38-39.

Background: The coronavirus disease pandemic of 2019 (COVID-19) has been associated with coagulopathy and an increased rate of thrombosis in adults. Medical practitioners have been prompted to consider prophylactic anticoagulation in special populations diagnosed with COVID-19. Patients with sickle cell disease (SCD) are predisposed to a hypercoagulable state. Despite the concern for development of venous thromboembolism (VTE) in these patients, there are no standardized guidelines for routine thromboprophylaxis in either adults or children with SCD. Thus, VTE management options are often extrapolated from guidelines for the general population. Method(s): A cross-sectional electronic survey was distributed to pediatric and adult hematology oncology practitioners through 7 SCD specific interest groups. Pediatric and adult practitioners were defined as those who medically manage patients 0-21 years of age and >21 years of age respectively. We examined responses to survey questions focused on routine thromboprophylaxis practices in children and adults with SCD prior to the pandemic and in patients with SCD admitted with COVID 19. Chi-square analyses or Fisher's exact tests for small samples were used to compare proportions as needed. Result(s): The survey was distributed to approximately 2,550 providers. Of 93 total responses, 14% (N=13) only treat patients >21yo; 38.7% (N=36) only treat patients 0-21yo and 47.3% (N=44) treat both. Nearly all adult practitioners (96.6%) would recommend pharmacologic prophylaxis, mechanical prophylaxis or both for hospitalized adults, but only 76% of pediatric treaters would recommend any prophylaxis (PPX) in hospitalized children (p<0.0001, Figure 1). Only 16% would recommend pharmacologic PPX only for patients 0-21yo, with 36% preferring non-pharmacologic only methods and 24% preferring both forms. In contrast, for the >21yo patients, 5 1% preferred to use both pharmacologic and non-pharmacologic PPX, 25% preferred pharmacologic alone, with just 5.3% preferring non-pharmacologic PPX only. Enoxaparin was the most frequently used anticoagulant among both pediatric and adult practitioners [78% vs 89% respectively] (Figure 2). Direct oral anticoagulants (DOACS) were infrequently recommended (3.8% for SCD children and 10.9% for adults); of these, rivaroxaban was used most often. The most common indication for starting thromboprophylaxis for both 0-21yo and >21yo patients was history of prior VTE [81% and 81% respectively], followed by hip replacement [57% and 75% respectively]. In patients admitted with COVID-19, prophylactic anticoagulation was recommended for adults by 94% of treaters. There was a significant increase in the use of prophylactic anticoagulation in children with COVID-19 vs. those without (84% vs. 40%; p=.0001). Figure 3 shows extended thromboprophylaxis practices upon discharge for COVID related admissions. Almost half of respondents (46%) would not recommend any thromboprophylaxis for children on discharge; 37% would recommend either 2-4 weeks or >4weeks of post-discharge PPX. The majority of adult providers would recommend discharge thromboprophylaxis for adults with 2 weeks post discharge (33%), and 2-4 weeks (30%) being the most common regimens. Conclusion(s): This pilot survey describes the thromboprophylaxis practices used for adult and pediatric SCD patients by specialty practitioners. In general, practitioners were likely to prescribe pharmacologic thromboprophylaxis for adults while mechanical PPX was preferred for children. However, in the high-risk setting of COVID-19 infection, pediatric practitioners would modify their practice to include S pharmacologic thromboprophylaxis. Interestingly, despite the long-term availability of direct oral anticoagulants in adults, and recent completion of pediatric studies, the most commonly used pharmacologic agent pre-COVID in adults and children was enoxaparin. Of note, this survey was conducted prior to release of the International Society on Thrombosis and Haemostasis guidelines regarding discharge PPX in COVID-19. (Spyr poulos AC, J Thromb Haemost, 2020) These results highlight the influence of COVID-19 on the use of pharmacologic thromboprophylaxis, specifically in the pediatric population. Due to frequent hospitalization, studies are needed to guide decision making surrounding VTE PPX for the adult and pediatric inpatient SCD population. (Figure Presnted).

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1. **[Hematology in the time of COVID-19]**  
   Gavillet Mathilde L'hematologie au temps du COVID-19. 2020;16:823-826.

The COVID-19 pandemic impacts the hematology practice. Intensive chemotherapies for high-grade lymphomas and acute leukemias, multiple myeloma treatments and most hematopoietic stem cell transplantations should be performed as usual. Low-grade lymphomas should only be treated when strictly indicated, maintenance can be postponed. Other myeloid neoplasia and their therapies cause imunosupression; dose adjustment is recommended but no brisk stopping. Sickle cell anemia patients are highly succeptible to severe COVID-19 course. Thrombocytopenia and procoagulant state are associated with severe courses of COVID-19, requiring an individualized therapy. No data indicate a risk of SARS-CoV-2 transmission through blood product transfusion.

[Available online at this link](https://www.knowledgeshare.nhs.uk/index.php?PageID=link_resolver&link=b0c2cd7724f299ab61a9db7d2cc14d3f)

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|  | **Source** | **Criteria** | **Results** |
| --- | --- | --- | --- |
| 1. | medline | exp coronavirus/ | 65995 |
| 2. | medline | exp Coronavirus Infections/ | 79552 |
| 3. | medline | ((corona\* or corono\*) adj1 (virus\* or viral\* or virinae\*)).ti,ab,kw,kf. | 3211 |
| 4. | medline | (coronavirus\* or coronovirus\* or coronavirinae\* or CoV).ti,ab,kw,kf. | 78269 |
| 5. | medline | ("2019-nCoV\*" or 2019nCoV\* or "19-nCoV\*" or 19nCoV\* or nCoV2019\* or "nCoV-2019\*" or nCoV19\* or "nCoV-19\*" or "COVID-19\*" or COVID19\* or "COVID-2019\*" or COVID2019\* or "HCoV-19\*" or HCoV19\* or "HCoV-2019\*" or HCoV2019\* or "2019 novel\*" or Ncov\* or "n-cov" or "SARS-CoV-2\*" or "SARSCoV-2\*" or "SARSCoV2\*" or "SARS-CoV2\*" or SARSCov19\* or "SARS-Cov19\*" or "SARSCov-19\*" or "SARS-Cov-19\*" or SARSCov2019\* or "SARS-Cov2019\*" or "SARSCov-2019\*" or "SARS-Cov-2019\*" or SARS2\* or "SARS-2\*" or SARScoronavirus2\* or "SARS-coronavirus-2\*" or "SARScoronavirus 2\*" or "SARS coronavirus2\*" or SARScoronovirus2\* or "SARS-coronovirus-2\*" or "SARScoronovirus 2\*" or "SARS coronovirus2\*" or covid).ti,ab,kw,kf. | 116487 |
| 6. | medline | (respiratory\* adj2 (symptom\* or disease\* or illness\* or condition\*) adj5 (Wuhan\* or Hubei\* or China\* or Chinese\* or Huanan\*)).ti,ab,kw,kf. | 334 |
| 7. | medline | (("seafood market\*" or "food market\*") adj10 (Wuhan\* or Hubei\* or China\* or Chinese\* or Huanan\*)).ti,ab,kw,kf. | 99 |
| 8. | medline | (pneumonia\* adj3 (Wuhan\* or Hubei\* or China\* or Chinese\* or Huanan\*)).ti,ab,kw,kf. | 603 |
| 9. | medline | ((outbreak\* or wildlife\* or pandemic\* or epidemic\*) adj1 (Wuhan\* or Hubei\* or China\* or Chinese\* or Huanan\*)).ti,ab,kw,kf. | 379 |
| 10. | medline | "severe acute respiratory syndrome\*".ti,ab,kw,kf. | 19298 |
| 11. | medline | or/1-10 | 144599 |
| 12. | medline | limit 11 to yr="2019 -Current" | 125180 |
| 13. | medline | Anemia, Sickle Cell/ | 21523 |
| 14. | medline | (HbS adj Disease).ti,ab. | 14 |
| 15. | medline | ("H?emoglobin S" adj Disease).ti,ab. | 22 |
| 16. | medline | ("Sickle Cell" adj (anemia or anaemia or disorder\* or disease or illness)).ti,ab,kw,kf. | 21591 |
| 17. | medline | 13 or 14 or 15 or 16 | 27681 |
| 18. | medline | Sickle Cell Trait/ | 2297 |
| 19. | medline | ("Sickle Cell" adj Trait\*).ti,ab,kw,kf. | 2177 |
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| 21. | medline | 17 or 20 | 28916 |
| 22. | medline | 12 and 21 | 90 |
| 23. | medline | (risk or probability or likelihood or prevalence or incidence).mp. | 3940935 |
| 24. | medline | 22 and 23 | 37 |
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| 2. | embase | exp Coronavirus infection/ | 24615 |
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| 5. | embase | (coronavirus\* or coronovirus\* or coronavirinae\* or CoV).ti,ab,kw. | 77622 |
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| 7. | embase | (respiratory\* adj2 (symptom\* or disease\* or illness\* or condition\*) adj5 (Wuhan\* or Hubei\* or China\* or Chinese\* or Huanan\*)).ti,ab,kw. | 404 |
| 8. | embase | (("seafood market\*" or "food market\*") adj10 (Wuhan\* or Hubei\* or China\* or Chinese\* or Huanan\*)).ti,ab,kw. | 108 |
| 9. | embase | (pneumonia\* adj3 (Wuhan\* or Hubei\* or China\* or Chinese\* or Huanan\*)).ti,ab,kw. | 652 |
| 10. | embase | ((outbreak\* or wildlife\* or pandemic\* or epidemic\*) adj1 (Wuhan\* or Hubei\* or China\* or Chinese\* or Huanan\*)).ti,ab,kw. | 175 |
| 11. | embase | "severe acute respiratory syndrome\*".ti,ab,kw. | 18892 |
| 12. | embase | or/1-11 | 150663 |
| 13. | embase | limit 12 to yr="2019 -Current" | 127231 |
| 14. | embase | \*sickle cell anemia/ | 24501 |
| 15. | embase | (HbS adj Disease).ti,ab. | 22 |
| 16. | embase | ("H?emoglobin S" adj Disease).ti,ab. | 25 |
| 17. | embase | ("Sickle Cell" adj (anemia or anaemia or disorder\* or disease or illness)).ti,ab,kw. | 31448 |
| 18. | embase | 14 or 15 or 16 or 17 | 34733 |
| 19. | embase | \*sickle cell trait/ | 1500 |
| 20. | embase | ("Sickle Cell" adj Trait\*).ti,ab,kw. | 2778 |
| 21. | embase | 19 or 20 | 3082 |
| 22. | embase | 18 or 21 | 36257 |
| 23. | embase | 13 and 22 | 140 |
| 24. | embase | (risk or probability or likelihood or prevalence or incidence).ti,ab. | 4870228 |
| 25. | embase | 23 and 24 | 53 |

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