

O impacto do COVID-19 nas pessoas com Doença Falciforme

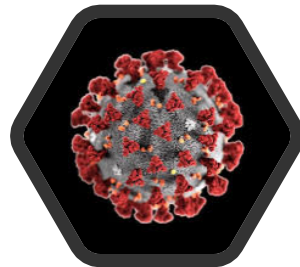
Comitê de Hematologia e Hemoterapia Pediátrica da ABHH

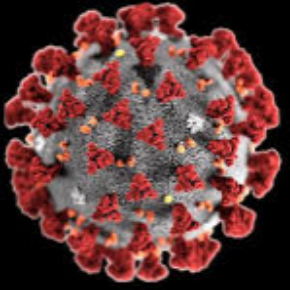
29 de Julho, 2020

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St. Jude Children's Research Hospital

COVID-19 and Sickle Cell Disease (SCD)

- SCD as an at-risk population
- Complications reported in SCD
- Results of an international registry for SCD patients
- My (limited) experience as of July 25, 2020
- Resources for patients from the SCDA





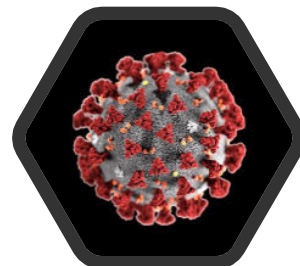
COVID-19: What is it? What are the symptoms?

- A new virus called **SARS-CoV-2**, or just coronavirus
- **SARS-CoV-2** causes Coronavirus Disease 2019 (COVID-19)
- Signs and Symptoms:
 - Flu-like symptoms: fatigue, muscle or body aches, headache, fever, chills, pharyngitis, runny nose
 - New loss of taste or smell
 - GI symptoms: Nausea, vomiting, diarrhea
 - Respiratory symptoms: cough, dyspnea, **pneumonia** (acute respiratory distress syndrome, **ARDS**)
 - **Hyperinflammatory state**, cytokine storm (multisystem hyperinflammatory syndrome, Kawasaki-like, in children), IL-6, TNF α
 - **Thrombosis (DVT), pulmonary embolism (PE)**
 - **Coagulopathy (elevated D-dimers, thrombocytopenia)**, with not much PT/PTT changes
 - Lymphopenia

In blue are complications of greatest mortality

COVID-19 could potentially cause **severe complications** in sickle cell disease (SCD) because...

- (1) Individuals living with SCD suffer from **hyperinflammation**
- (2) Individuals with SCD are at higher risk of **deep venous thrombosis**
- (3) Individuals with SCD are **immunocompromised** due to auto-infarction of their spleen or surgical splenectomy
- (4) Individuals with SCD often have **comorbidities and secondary organ dysfunction** that can put them at risk of significant morbidity and or mortality



CDC considers **SCD** a high-risk condition for **severe illness** from COVID-19

1. Cancer
2. Chronic kidney disease
3. Chronic obstructive pulmonary disease
4. Immunocompromised state from solid organ transplant
5. Obesity (BMI of 30 or higher)
6. Serious heart disease (heart failure, coronary artery disease, cardiomyopathy)
- 7. Sickle cell disease**
8. Type 2 diabetes mellitus



Centers for Disease Control and Prevention
CDC 24/7: Saving Lives, Protecting People™

www.cdc.gov

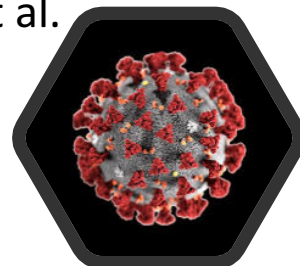
Current literature of COVID-19 and SCD

33 publications as of July 24, 2020

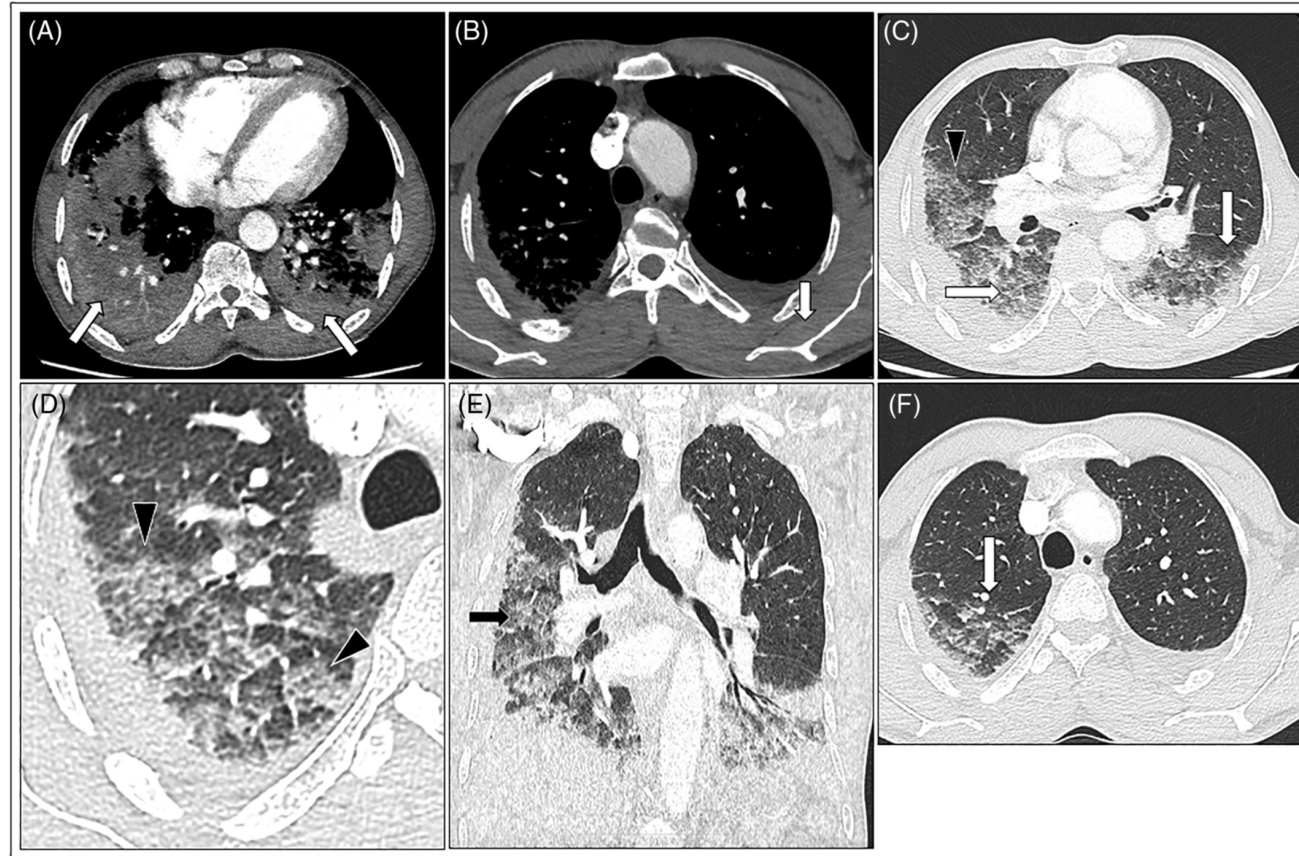
- 134 cases (children and adults)
- **Most common presentation:** Pain, ACS. Pain (VOC) may be the only presenting symptom (triggered by COVID-19?), sometimes no fever (Al-Hebshi A, PBC 2020 Jul 10)
- Pulmonary embolism and severe ACS
- **Pregnancy:** trigger severe VOC, ACS (Justino C. Hematol Transfus Cell Ther . 2020 Jun 18)
- **Labs:** worse anemia, lymphopenia, ↑ cytokine levels (IL-6), ↑ CRP, ↑ D-dimers
- **ICU admission and Mortality:** more common among older patients and with severe disease (**age >45 years** - Arlet et al. Lancet Haematol. 2020 Jun 18;S2352)

Treatment

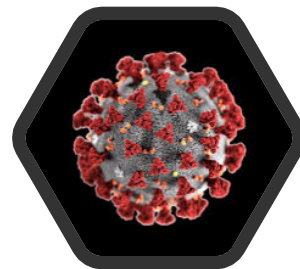
- Severe ACS treatment: **exchange transfusion**, dexamethasone (watch for rebound pain/ACS), Antibiotics
- Antiviral Treatment: **remdesivir**, hydroxychloroquine
- Cytokine storm: **anakinra** (human interleukin-1 receptor antagonist, IL-1Ra -(Appiah-Kubi A. et al, Br J Haematol 2020 Jul 11), **tocilizumab** (anti-human IL-6 receptor monoclonal antibody)



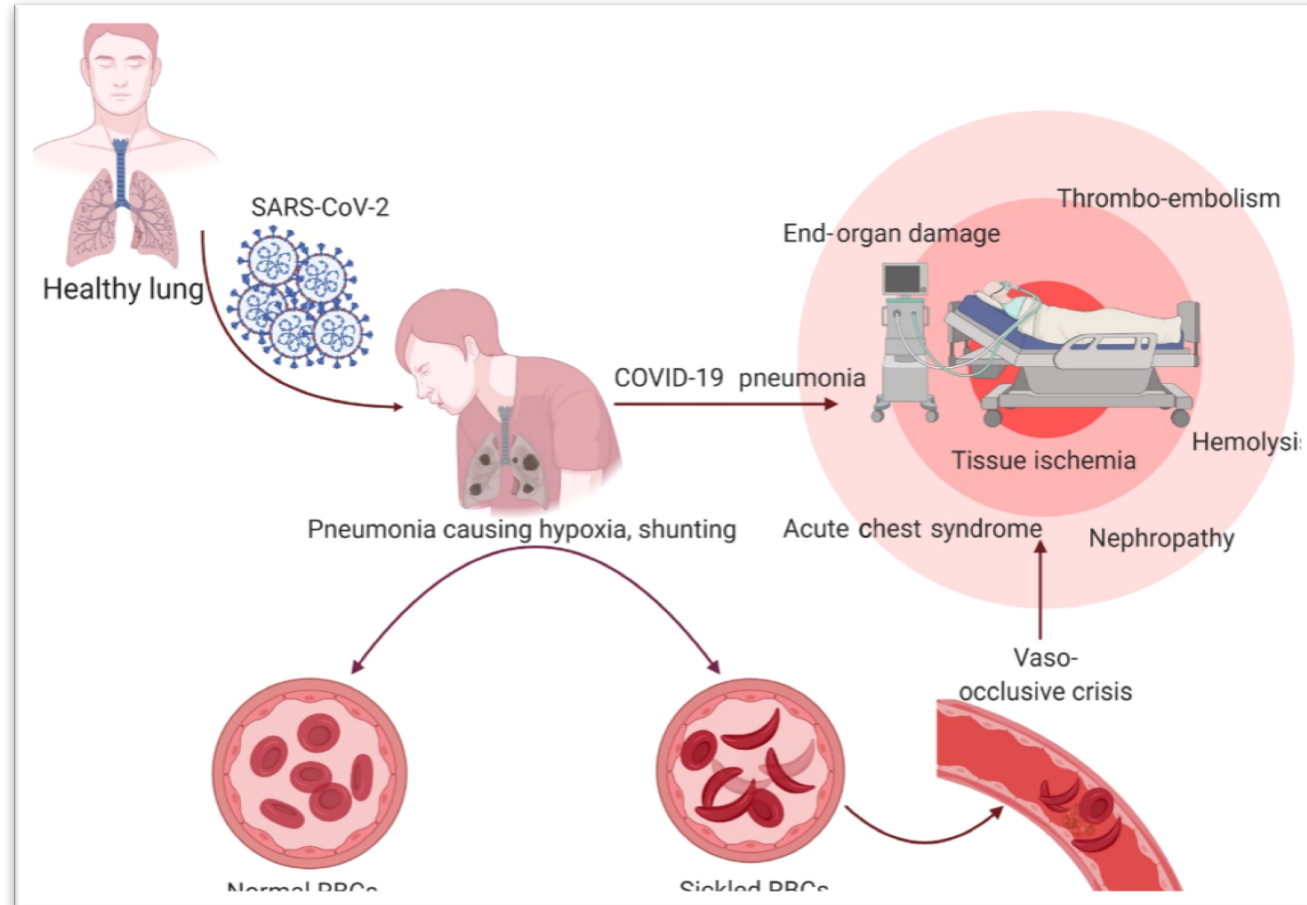
Tocilizumab (anti-human IL-6 receptor monoclonal antibody) 8 mg/kg for severe ACS



45 yo HbSS (images at presentation) De Luna G. et al. Am J Hematol . 2020 Jul;95(7):876-878.
16 yo HbSS with ACS and PE and very high IL6. Odievre MH. et al. Am J Hematol. 2020 Aug;95(8):E192-E194.



Complications of COVID-19 in people with SCD



Sahu et al., British Journal of Haematology, Volume: 190, Issue: 2, Pages: e86-e89, 23 May 2020.

International Registry for COVID-19 in SCD



Reporting of COVID-19 cases in SCD globally

This form is to be completed by a health care professional caring for a patient with sickle cell disease and documented coronavirus (COVID 19).

Please report only confirmed COVID-19 cases, and report after sufficient time has passed to observe the disease course through resolution of acute illness and/or death.

Fields marked with a red asterisk (*) are required.

If you have any questions, please contact us at: covid.sicklecell@mcw.edu.

Reporter Information

Name of reporter *

Email address of reporter *

Name of physician providing care for Sickle Cell disease *

Name of center/practice providing care for Sickle Cell disease *

Patient Information

Age, in years *

As of July 17, 2020



Count of Cases Reported

260

Age Summary

26.83

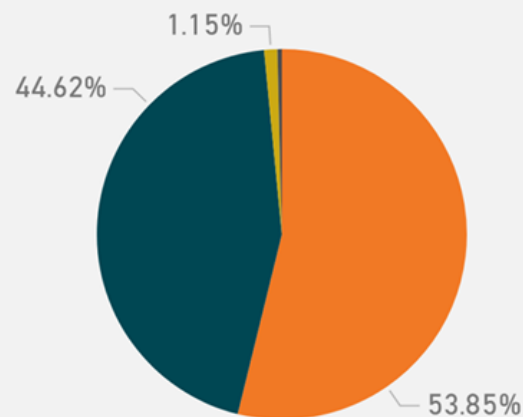
Average Age

15.12

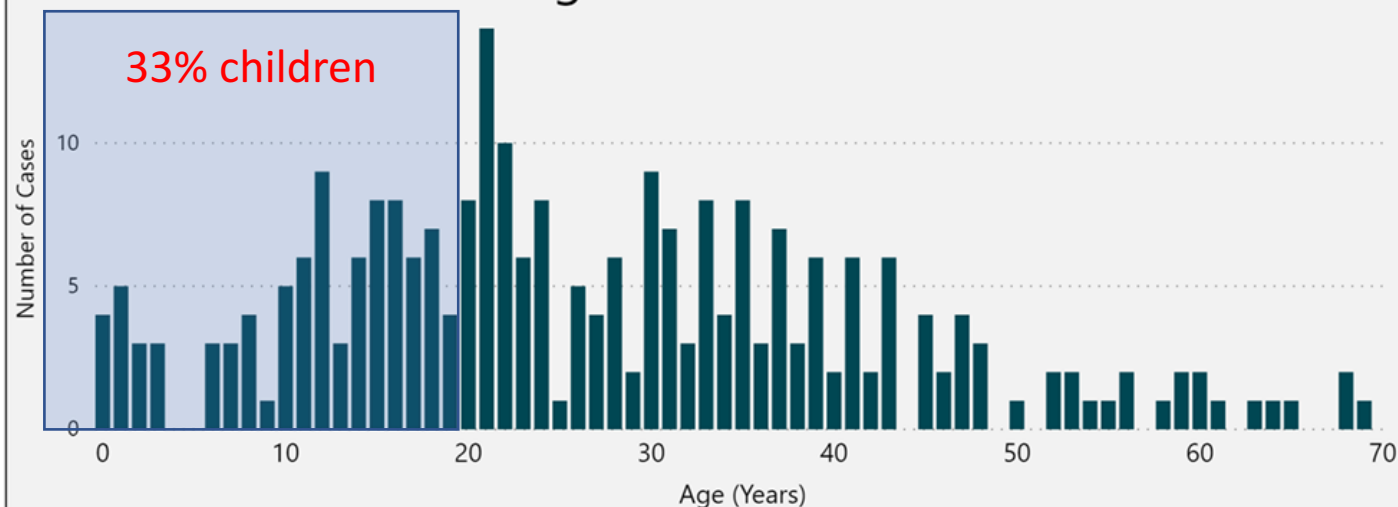
Standard Deviation

Sex

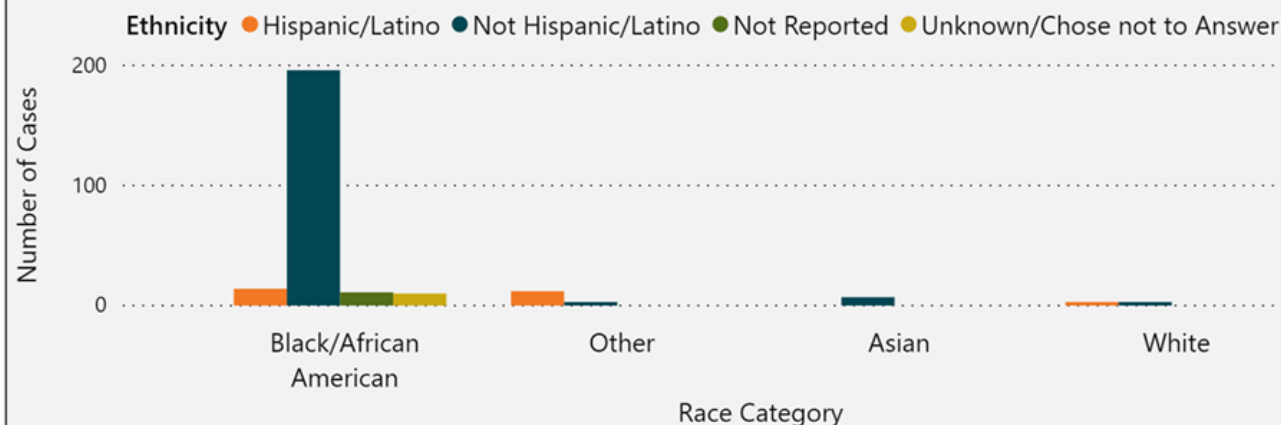
Female Male No Answer Other



Age Distribution

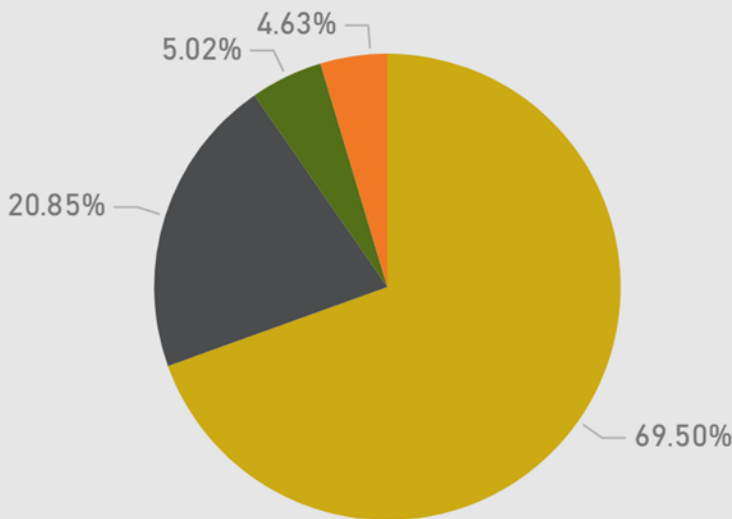


Race & Ethnicity Distribution



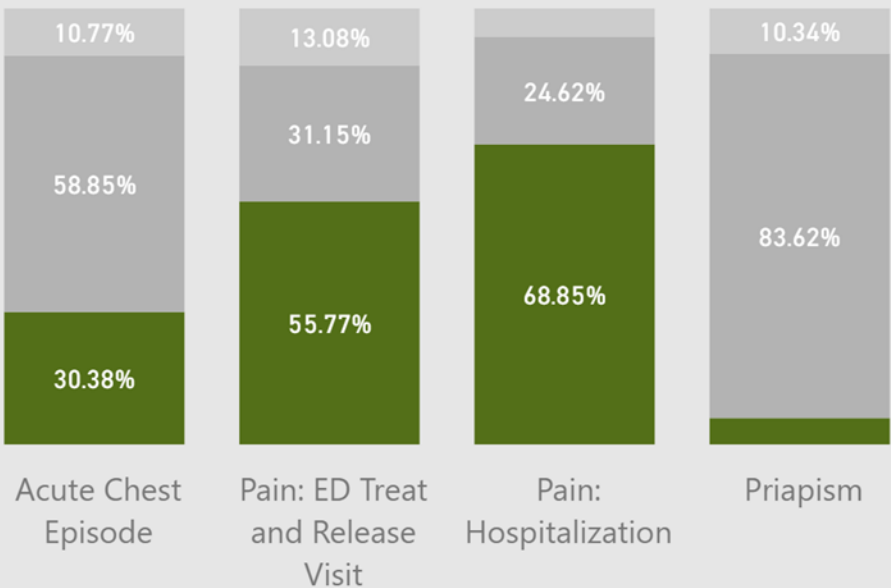
Sickle Cell Disease Type (% of Total)

● Hgb SS disease ● Hgb SC disease ● Hgb S beta + thalassemia ● Hgb S beta zero thalassemia



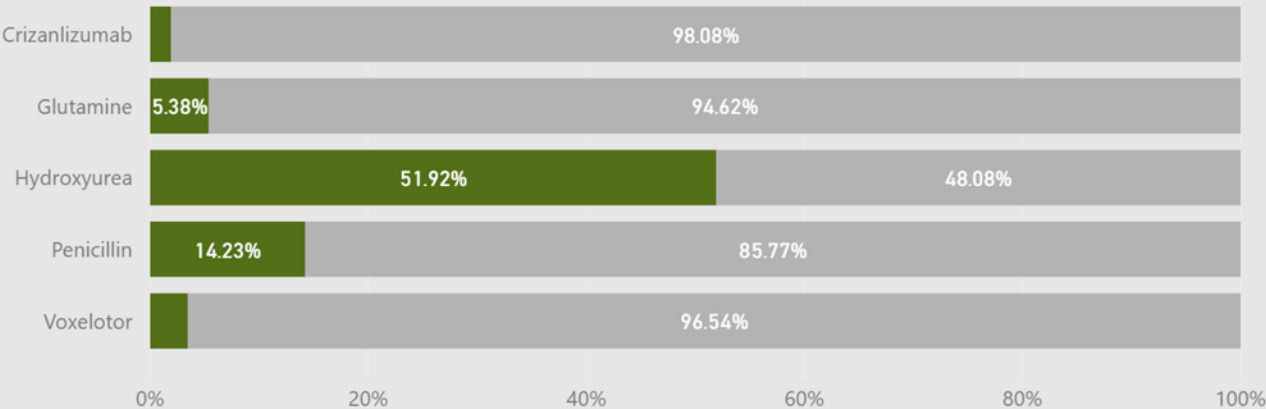
SCD Symptoms in Previous 3 Years

● Yes ● No ● Unknown



SCD Home Medication Use

Med Status ● Taking ● Not Taking



Average

Standard Deviation

Pain: Hospitalization

7.99

9.11

Pain: Treat and Release ED Visit

8.76

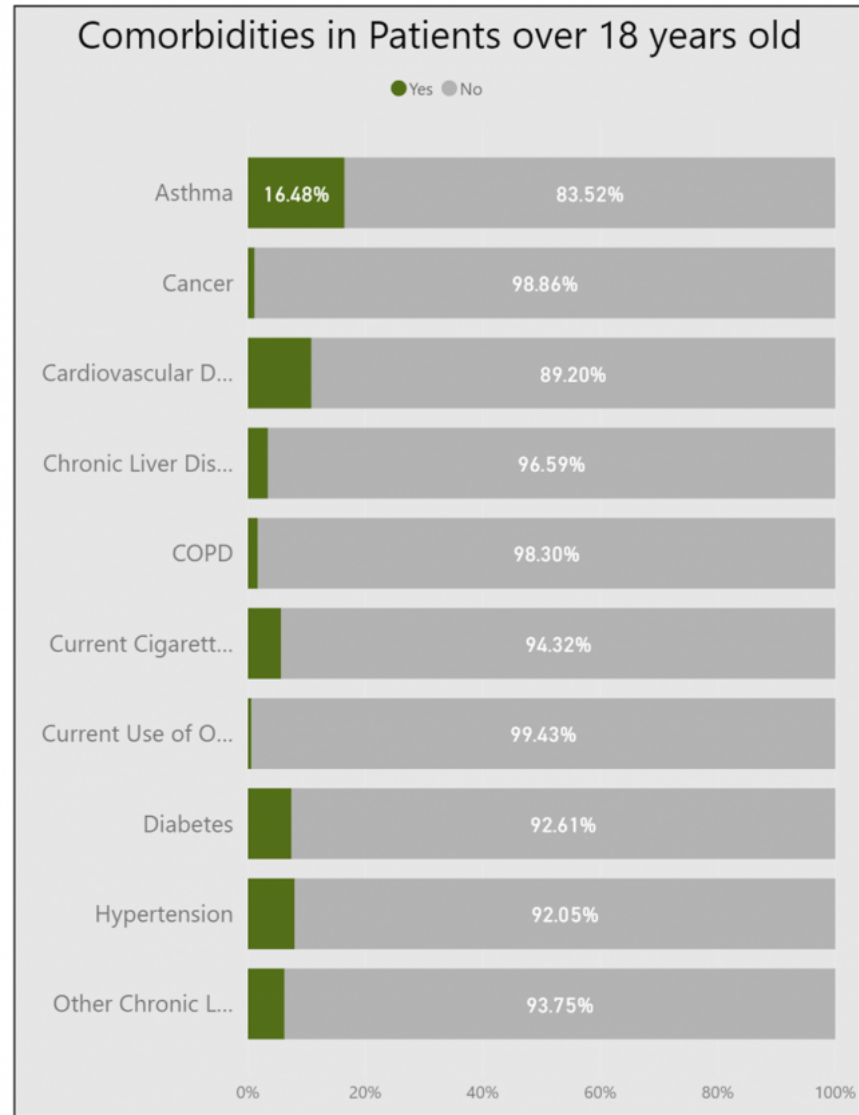
16.34

Acute Chest Episodes

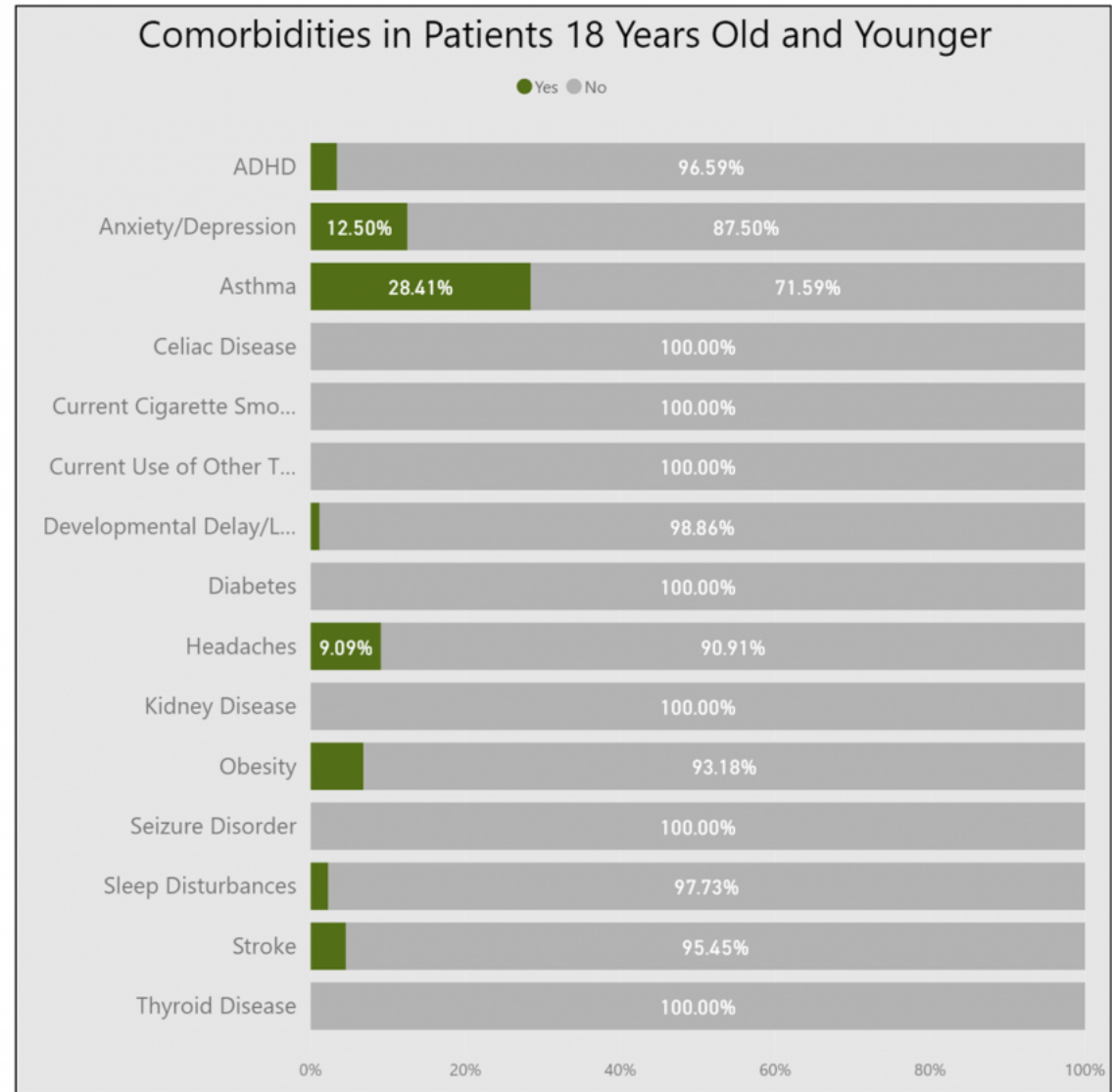
2

2

Adults

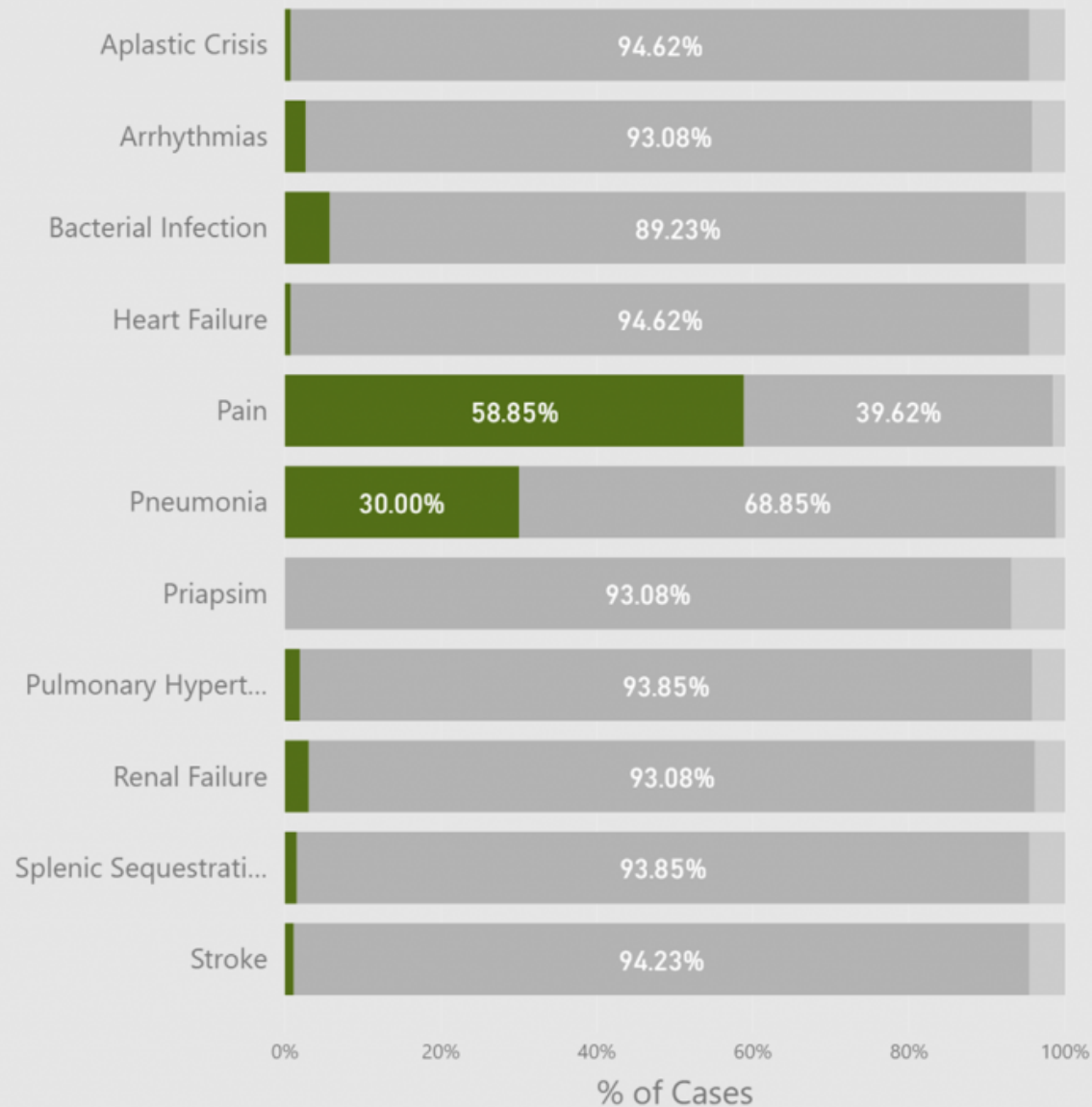


Children



Symptoms during COVID-19 Infection

● Yes ● No ● Unknown



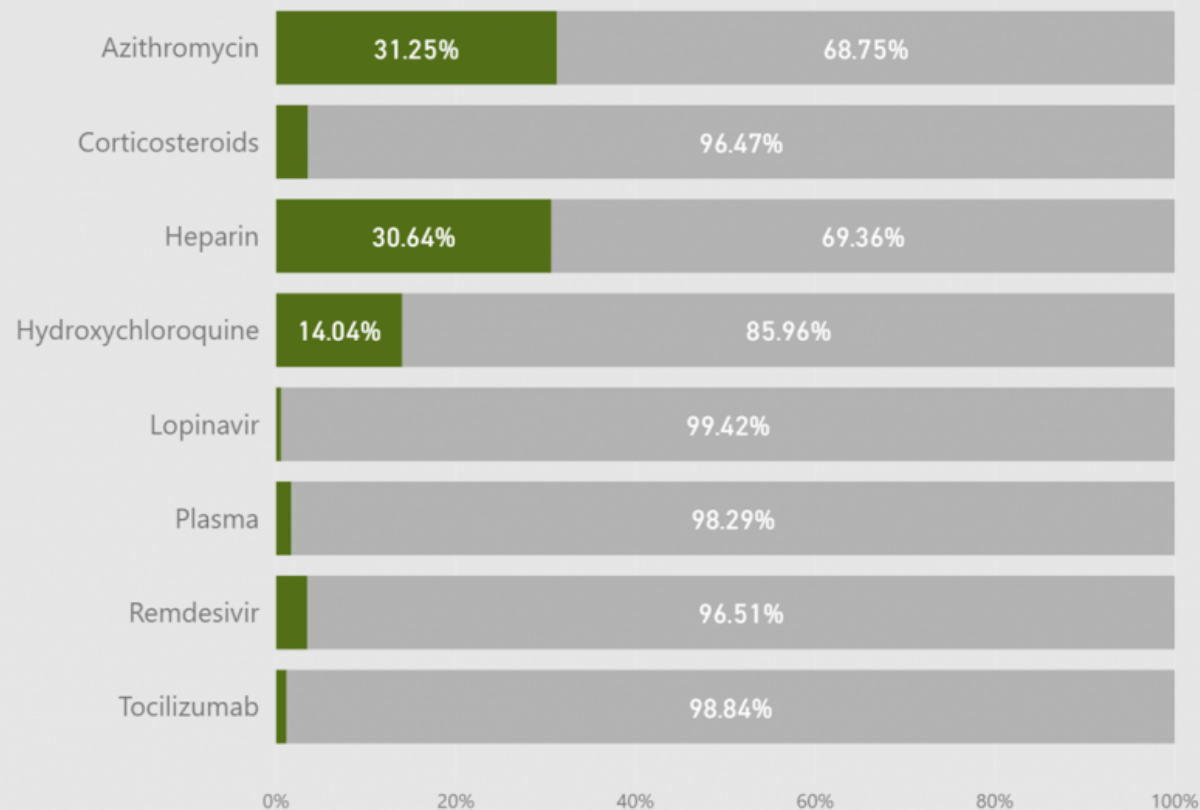
What are the most common complications of COVID-19 in sickle cell disease?

Pain and Acute Chest Syndrome are the most common complications



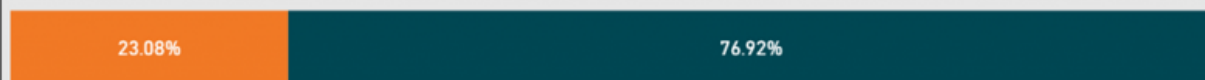
COVID-19 Treatments - Medication Use

● Yes ● No



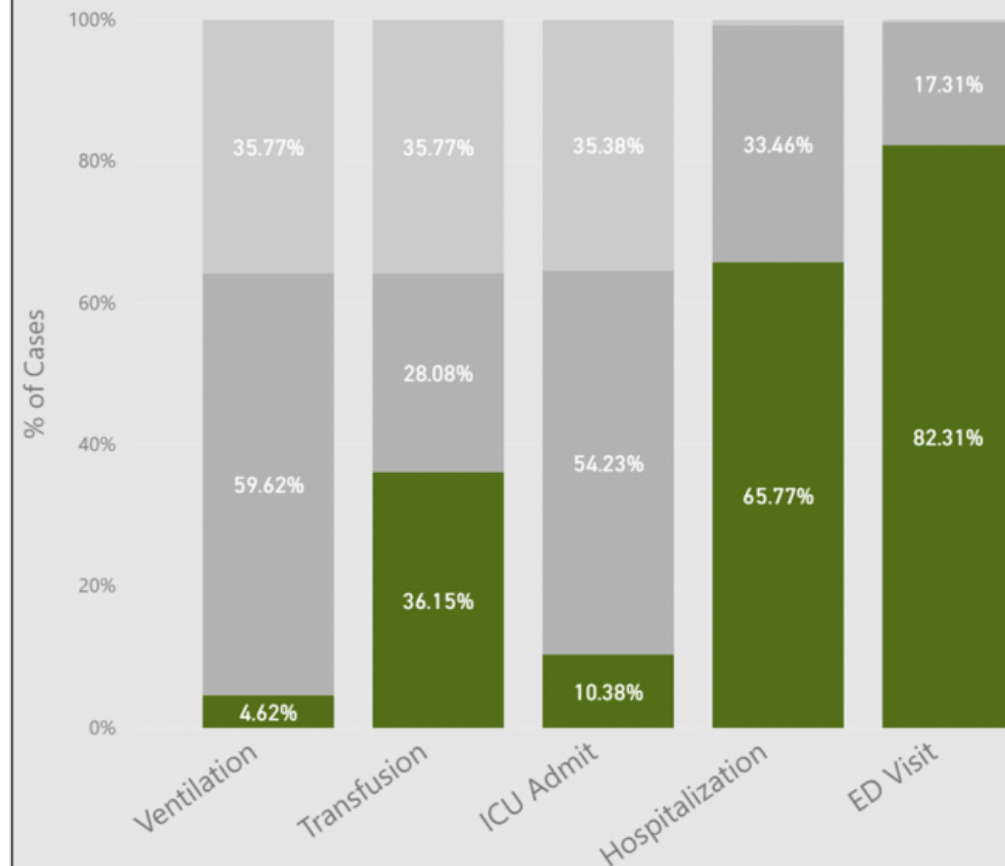
Heparin Use

● Therapeutic ● Preventative



COVID-19 Interventions

● Yes ● No ● Not Reported



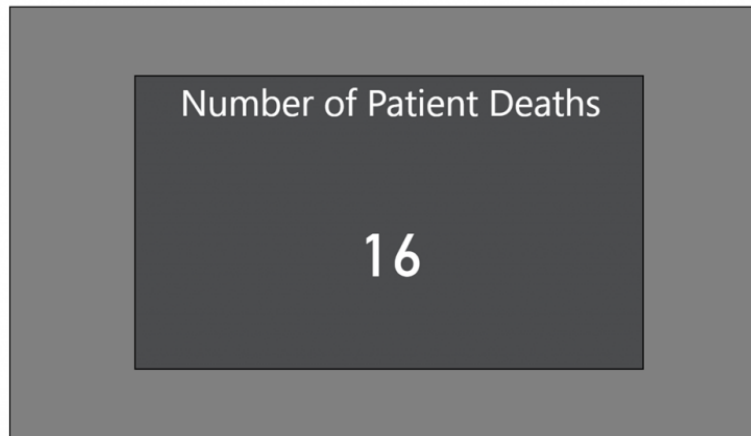
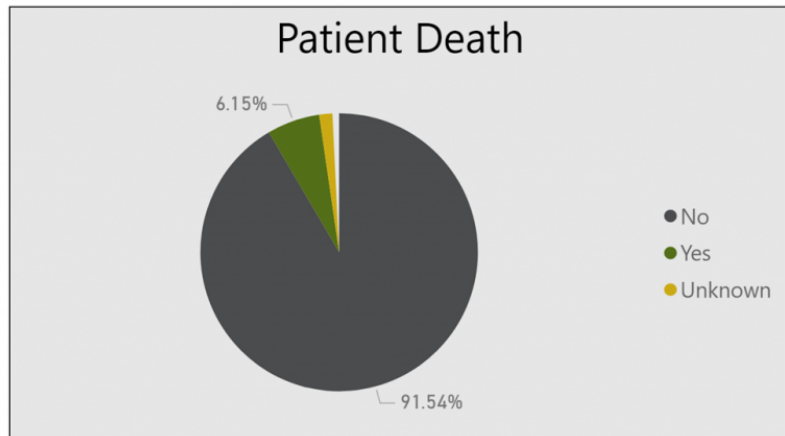
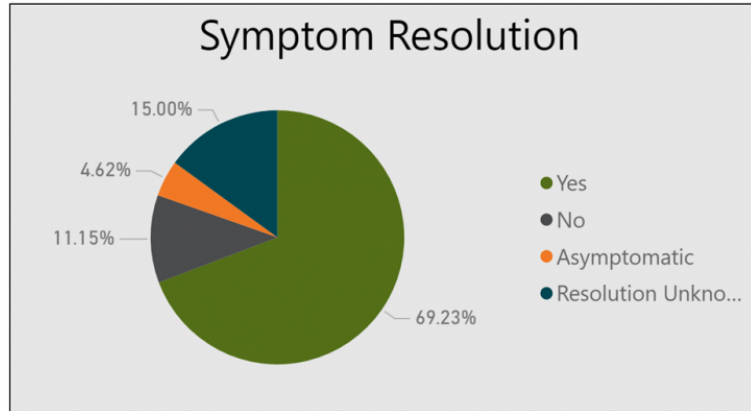
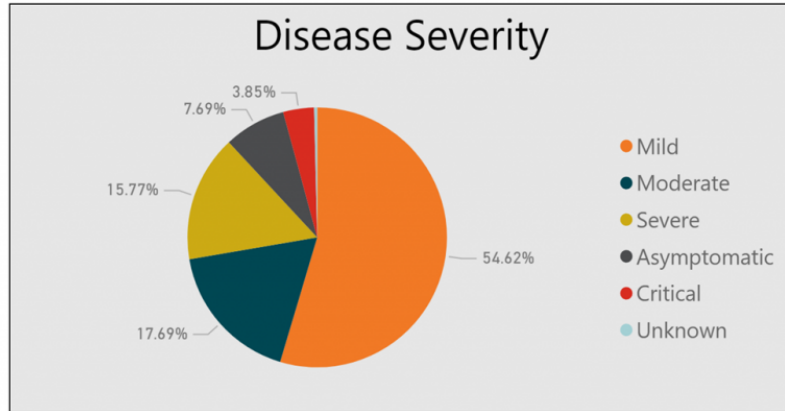
Average
Hospital LOS

7.83

Transfusion Type

● Exchange Transfusion ● Simple Transfusion





What's the **mortality** from COVID-19?

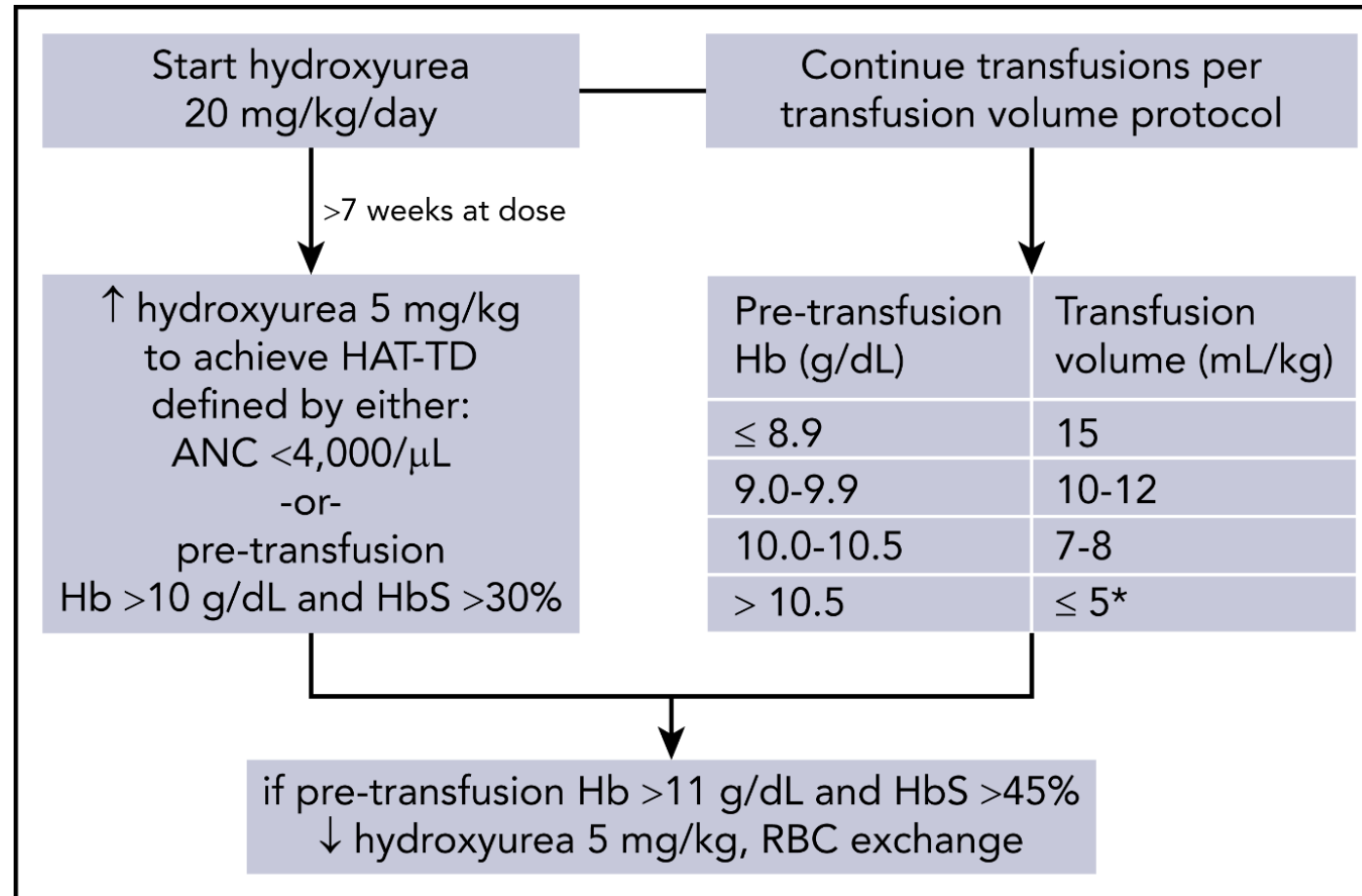
6% of people with SCD died of COVID-19 (1 child)

Compared general population:

US 3.5%

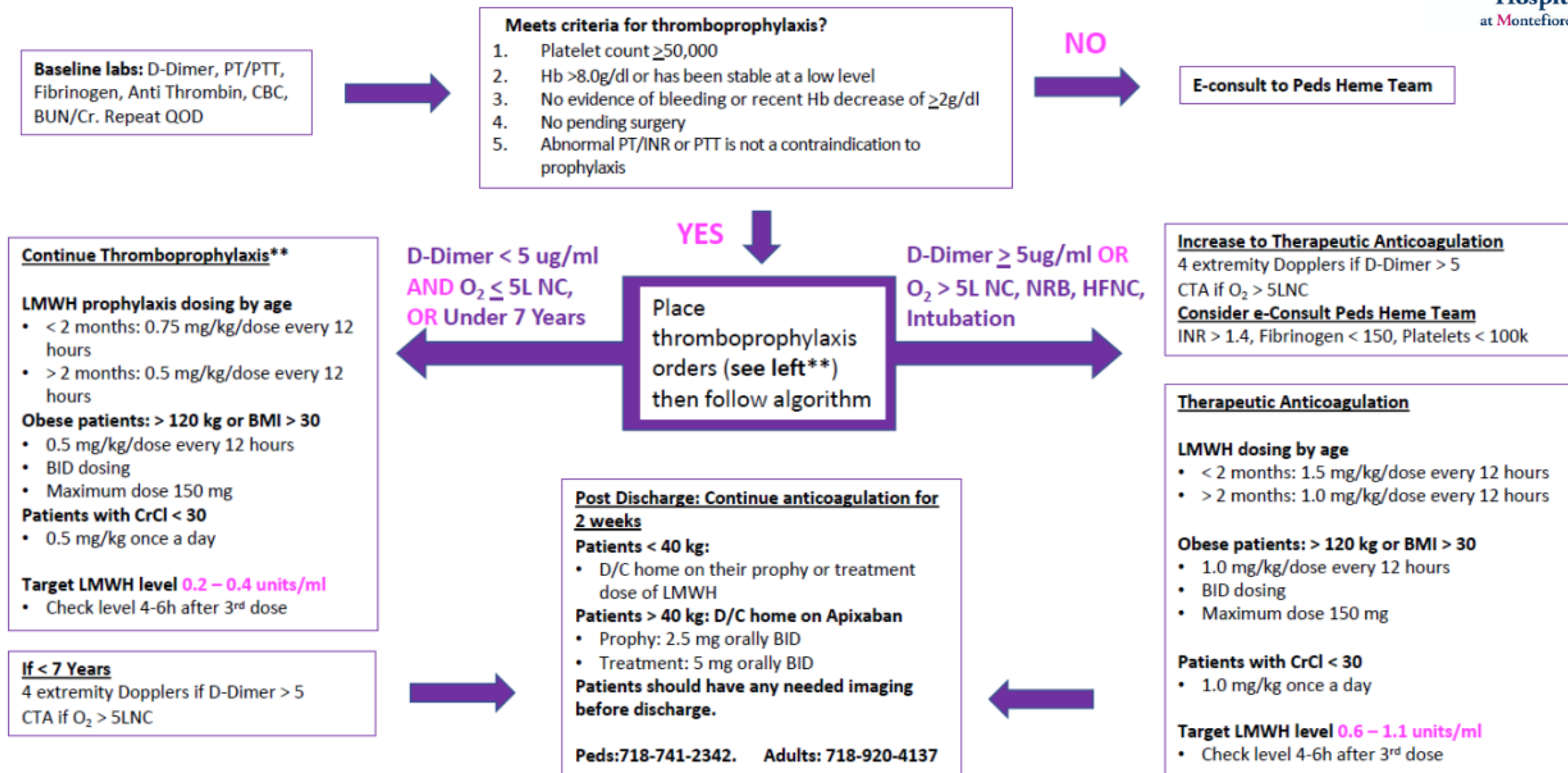
Brazil 3.7%

Combination dose-escalated hydroxyurea and transfusion: an approach to conserve blood during the COVID-19 pandemic



Nickel R. et al. Blood, 2020.

CHAM Anticoagulation Protocol for COVID Positive Patients 7 Years and Older



My experience at St. Jude

17 children

- Age ranged 9 months to 17 years
- Most mild symptoms
- 3 admissions (1 to the ICU)
- Most common signs/symptoms:
 - Fever
 - Pain crisis
 - Headache
 - ACS
 - Pharyngitis

Single case to the ICU:

- Respiratory distress 6 days after onset of COVID-19
- Rapid deterioration once tachypnea ensued
- Exchange transfusion given on D1
- Dexamethasone 6 mg IV/day (gave only 5 days owing to risk of rebound pain/ACS)
- Remdesivir ordered by not given due to improvement with exchange transfusion
- Enoxaparin (D-dimer >20 mg/mL)

Resources for patients
with SCD

SCDAA
(sicklecelldisease.org)

Lifestyle Adjustments

During this time, you may feel anxious. However, **you do have control over what you do!**

Helpful Tips!

- Meditate*
- Keep a Routine
- Read a Book
- Exercise at Home
- Listen to Music
- Video Chat
- Call Your Friends

REMEMBER
KEEP A SAFE DISTANCE...
SIX FEET

SCDAA is committed to helping families get through this health crisis.

Sickle Cell Disease Association of America, Inc.
7240 Parkway Drive, Suite 180
Hanover, MD 21076
admin@sicklecelldisease.org
410.528.1555

* There are apps to help you learn to meditate.

Social Adjustments & Response to COVID-19

Advice for People with Sickle Cell Disease & SCD Families

COVID-19, also known as Coronavirus, is a new respiratory illness that can be very serious.

We all need to do our part to protect ourselves and others, so the virus does not spread.

WHAT CAN YOU DO?

WEAR A MASK – When in public to reduce the spread of the virus.



PHYSICAL DISTANCING – Avoid close contact with others to reduce the spread of the virus.



FREQUENT HAND WASHING – At least 20 seconds with soap and water.



CALL YOUR PROVIDER – If you have symptoms.



SYMPTOMS

MILD

Shortness of breath
Dry Cough
Mild Fever
Change in taste or smell sensations

SEVERE

High Fever
Severe Respiratory Disease
Pneumonia



AVOID

- Group Gatherings
- Sleepovers
- Playdates
- Visitors to your Home
- Constantly Watching the News



CAUTION

Visit Grocery Store
Get Take Out
Pick Up Medications
Mass Transit Systems
(keep your distance, avoid touching surfaces)
Traveling



SAFE

- Yard Work
- Play in your Yard
- Clean out a Closet
- Read a Good Book
- Listen to Music
- Cook a Meal
- Family Game Night
- Group Video Chats
- Stream a Favorite Show
- Check on a Friend
(from a distance)
- Check on Elderly Neighbor
(from a distance)
- Virtual Religious Service



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Hemoglobin disorders such as sickle cell disease and thalassemia

Having sickle cell disease (SCD) increases your risk for severe illness from COVID-19. Having other hemoglobin disorders, like thalassemia, may increase your risk for severe illness from COVID-19.

Actions to take

- Ask your healthcare provider about telemedicine or remote healthcare visits, and know [when to go to the emergency department](#).
- Work with your healthcare provider to manage [medicines and therapies](#) for your disorder (including hydroxyurea, chelation therapy, blood transfusions, and prescriptions for pain management) and any other health condition you may have (such as diabetes, high blood pressure, and arthritis).
- **If you don't have a healthcare provider**, contact your nearest [community health center](#)  or [health department](#).
- Try to prevent vaso-occlusive episodes or pain crises by [avoiding possible triggers](#).
- Review CDC's [healthy living with SCD guide](#) or our [healthy living with thalassemia guide](#) for tips to help you stay healthy.
- Find [SCD resources](#) and [thalassemia resources](#) to help navigate care and increase knowledge and awareness of SCD and thalassemia.
- Let friends and family know about the need for [healthy blood donors](#).



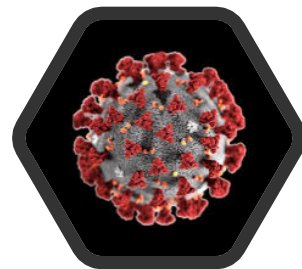
Highlights of management of COVID-19 in SCD

1. Most cases do well, those who with worse course usually adults
 2. Pain and ACS main complications
 3. ACS → exchange transfusion quickly, tocilizumab may also have benefit
 4. Prophylactic anticoagulation if hospitalized, monitor D-dimer
 5. Add hydroxyurea to reduce blood utilization
 6. Cytokine storm → consider tocilizumab or anakinra
 7. Pregnancy → expect more severe pain events and other complications
-



Obrigada!

Extra slides



What should I do about **School** this fall?

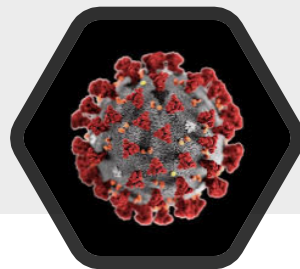
- Many schools offering choice: **distance learning** (online) versus **in-person**
- Chose based on:
 - What works best with your schedule
 - If you chose in person, ask how school is preparing: how many kids in the classroom? How will the school be cleaned? Will kids and school staff be required to wear a mask? Will kids be kept in small cohorts? Will kids and staff have breaks for hand washing frequently?

There is no right answer and whatever you chose will be the right thing for your child!



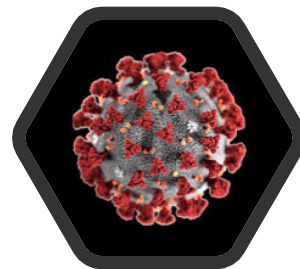
How do we treat infection with COVID-19 in a person with sickle cell disease?

- **Steroids** seem to be very helpful, but may cause **side effects in people with sickle cell disease** (rebound pain – about 7 days after using it)
- **Hydroxychloroquine** (anti-malarial): does not work to prevent infection if you got in touch with a person with sickle cell disease (not a prophylactic medicine). Probably not a good medicine too treat COVID-19 either.
- **Remdesivir** (antiviral): does not reduce mortality from COVID-19, but shortens duration of disease



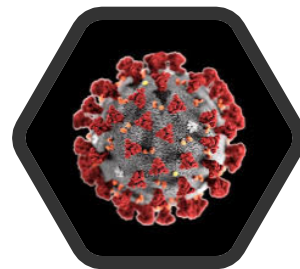
How can do you prevent COVID-19 in a person with Sickle Cell Disease?

- Wash your hands frequently for 20 seconds
- **Use a hand sanitizer that contains at least 60% alcohol**
- Wear a cloth mask
- Distance from others by at least 6 feet (avoid crowds)
- If someone in your house has COVID-19, keep separate rooms
- Cover coughs and sneezes
- Clean and disinfect surfaces (door knobs, light switches, phones, etc.)
 - bleach and water is fine

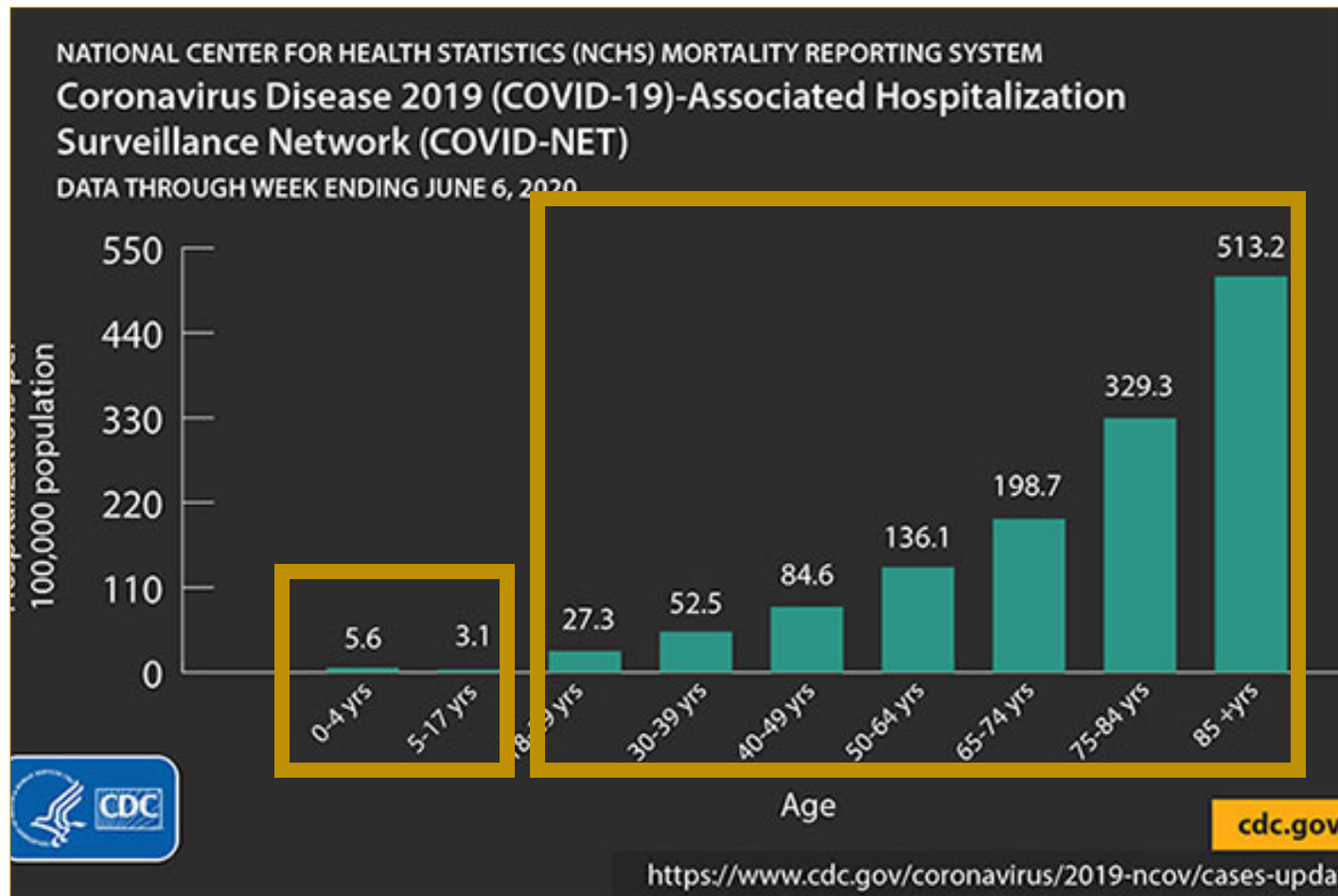


What about a vaccine for COVID-19?

- **Vaccine:** none yet
- **>100** being tested
- **Moderna (Cambridge, MA)** and **University of Oxford, UK** are the front runners (in phase II and III clinical trials)
- Both are **genetically engineered** vaccines: tells your body to make parts of the coronavirus, and your immune system recognizes and produces antibodies



But, **age** is one of the most important factors:



The older you are, the more likely it is to be hospitalized (admitted)