



ARISE

African Research And Innovative  
Initiative For Sickle Cell Education

# SICKLE CELL DISORDER AND COVID-19: Symptom overlap

By

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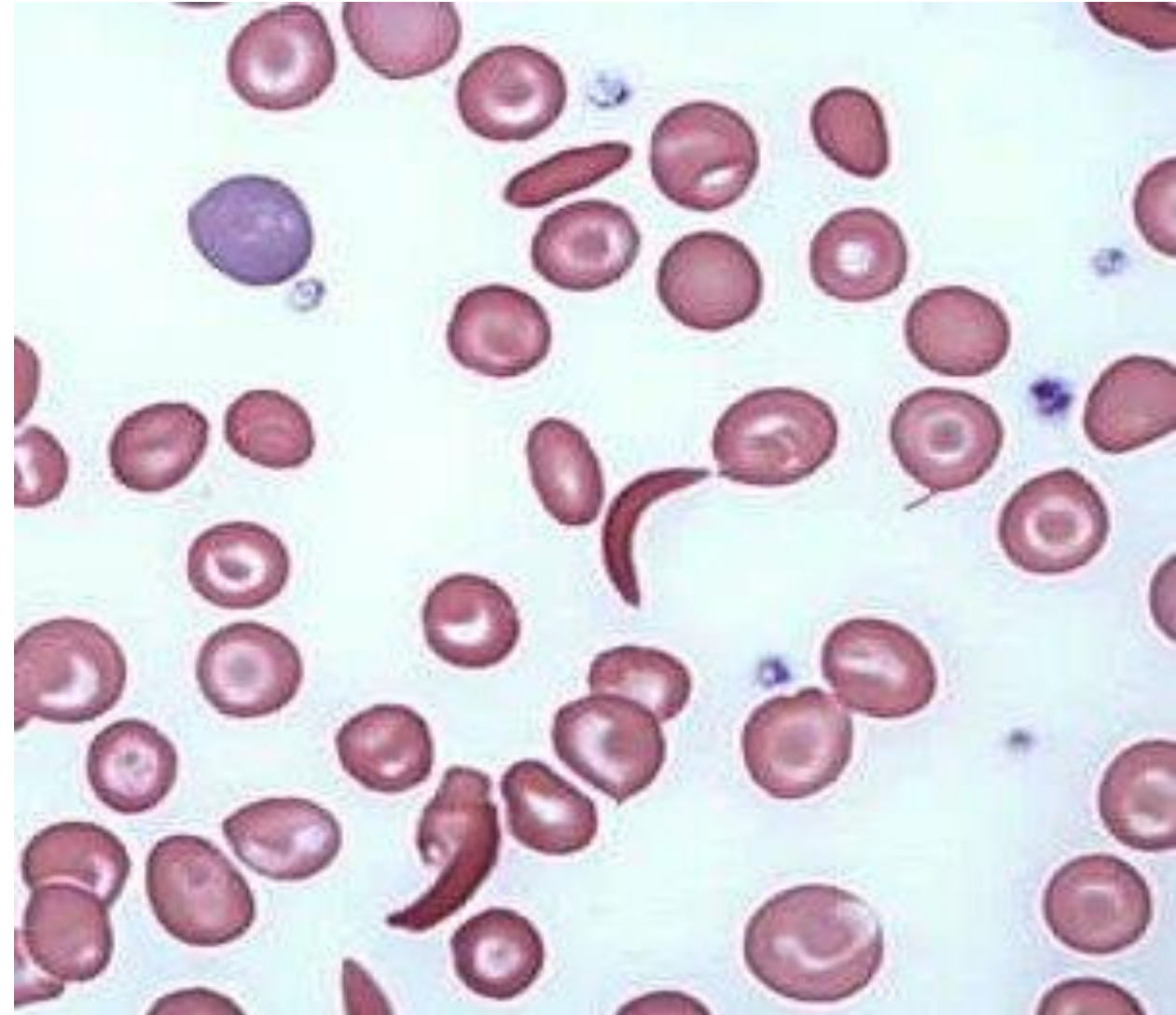
# OUTLINE

- What is sickle cell disorder?
- Pathophysiology; what goes wrong?
- SCD and COVID-19
- Acute chest syndrome Vs. COVID-19
- Management and preventive strategies
- Conclusion
- Resources
- Appreciation



# What is sickle cell disorder?

- Group of disorders characterized by the inheritance of 2 unusual Hb genes, 1 of which is the sickle gene (Hb S)
- SCDs include; SC, SD, S $\beta$ -thal, SS
- SCA = Hb SS

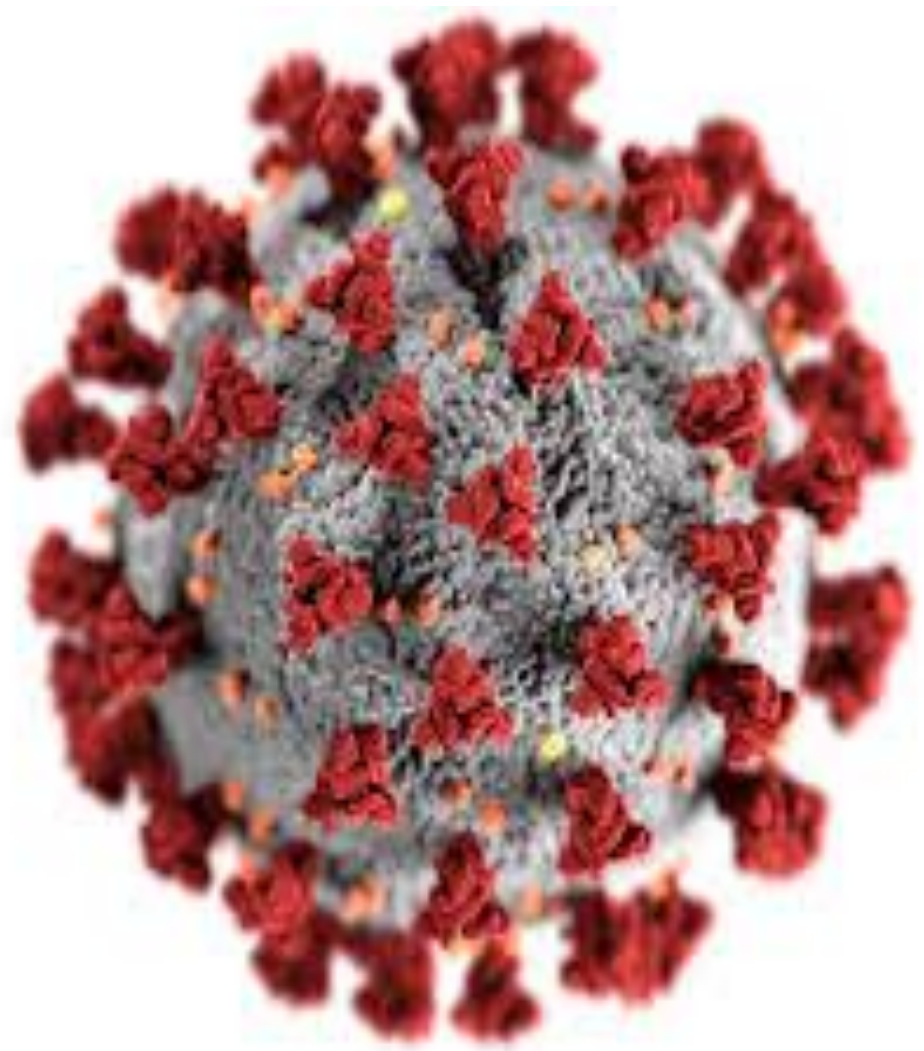


# Pathophysiology: What goes wrong?

- Hb undergoes physicochemical changes when oxygen levels are reduced → sickle shaped cells
- There's a complex interplay between blood vessels, red blood cells, white blood cells, platelets and special proteins in the blood
- Spectrum of events; haemolysis to ischaemia
- Underlying mechanism of clinical features
- Complicated by acute events e.g. anaemia, acute chest syndrome, priapism etc.
- Chronic complications e.g. avascular necrosis of the head of the femur (AVN), chronic osteomyelitis, leg ulcers etc.



- Respiratory illness caused by SARS CoV-2
- 1<sup>st</sup> documented in Wuhan, China
- High transmissibility, CFR 2.6%
- 8,468,727 cases, 451,980 deaths globally and 17,735 cases, 469 deaths in Nigeria (18/06/20 12.36 GMT)
- Suspected case:  
Cough ± fever (or history of fever in the last 2 weeks) + ≥1 of chills, body pain, headache, sore throat, recent loss of taste or smell, difficulty in breathing, diarrhoea/abdominal pain, runny nose, fatigue NCDC



# SCD & COVID-19

- Lower immunity than general population, disruption in routine care
- Hb SS identified as 'clinically extremely vulnerable group' in the UK, 'vulnerable' in the US
- Increased risk of infections including COVID-19
- May develop ACS (from other causes, may co-exist with COVID-19) leading to diagnostic conundrum





# Acute Chest Syndrome

- Potentially fatal complication
- ‘finding of a new pulmonary infiltrate involving at least one complete lung segment that was consistent with the presence of alveolar consolidation, but excluding atelectasis. .... **chest pain**, a **temperature** of more than 38.5°C, tachypnea, wheezing, or **cough**’

## Atelectasis

- Pain due to rib and vertebral infarction
- Reduced respiratory drive and cough

## Fat embolism

- Bone marrow infarction
- Release of phospholipase A2

Acute chest syndrome

## True thromboembolism

- Activated endothelium
- Reduced red cell flexibility
- Increased red cell adherence
- Vaso-occlusion

## Infection

- *Chlamydia pneumoniae*
- *Mycoplasma pneumoniae/hominis*
- *Staphylococcus aureus*
- *Streptococcus pneumoniae*
- Respiratory syncytial virus
- Parvovirus
- Rhinovirus
- Influenza and parainfluenza virus



# ACS Vs COVID-19

	ACS	COVID-19
<b>Epidemiology</b>	Any age, ethnicity	Elderly, vulnerable populations, ethnicity
<b>Aetiology</b>	Atypical organisms, fat infiltrates	SARS-CoV-2
<b>Smoking,</b>	Increased risk	Increased risk
<b>Clinical features</b>	Cough, fever, difficulty in breathing	Cough, fever, difficulty in breathing, loss of smell/taste, sore throat
<b>Radiological findings</b>	Localized infiltrates	Diffuse ground glass appearance
<b>Transfusion</b>	Exchange/top up, standard of care	Outcome of transfusion unclear





# Management and preventive strategies



**Sickle Cell Disease and COVID-19:  
A guide to decrease burden and minimize morbidity in the context of emergency lockdown  
in Kaduna State.**

*SCD and Haematology management team  
(Adapted from Medical and Research Advisory Committee Sickle Cell Disease Association of  
America for BDTH and affiliated clinics)*



# Management and preventive strategies

- Practice **shielding**

- Well ventilated areas, outdoors (strict conditions)
- Avoid non-residential individuals, limit to 1 if alone
- Sleep in separate beds
- Individual towels, toilets, bathrooms etc.
- Assisted shopping, medication fill-ups etc.
- NO PARTIES!!
- NO CONTACT with individuals with symptoms of COVID-19
- Can be implemented in the workplace

- Hand and respiratory hygiene



# Management and preventive strategies

- Maximise tele/e-medicine/e-consultation options
- Home use of thermometers
- Routine medication including hydroxyurea
- PCA as indicated, liaise with clinician & pharmacy for controlled drugs
- Triage according to protocols if COVID-19 symptoms are present
- If COVID-19 positive;
  - High index of suspicion for ACS, institute standard treatment
  - Evaluate for cardiopulmonary co-morbidities e.g. PH, asthma
  - Review other meds like ACEIs
  - Transfusion as needed



# Conclusion

- SCD constitutes a high risk group for infections including COVID-19
- Adjustments by patients, health care systems required to ameliorate risk
- Low threshold for action when SCD patients have respiratory symptoms suggestive of COVID-19
- Institute protocols for standards of care to improve outcomes



# Resources

- <https://www.gov.uk/government/publications/guidance-on-shielding-and-protecting-extremely-vulnerable-persons-from-covid-19/guidance-on-shielding-and-protecting-extremely-vulnerable-persons-from-covid-19#handwashing-and-respiratory-hygiene>
- <https://sicklecellanemianews.com/information-about-covid-19-sickle-cell-disease/>
- <https://sicklecellanemianews.com/2020/05/07/sickle-cell-association-issues-covid-19-guidelines-for-scd-patients-us-reopening/>
- [https://covid19.ncdc.gov.ng/media/files/Community Case Definition PDF.pdf](https://covid19.ncdc.gov.ng/media/files/Community_Case_Definition_PDF.pdf)
- Yusuf BJ, Abba AA, Tasiu M. Acute chest syndrome. Sub-Saharan Afr J Med [serial online] 2014 [cited 2020 Jun 18];1:111-8. Available from: <http://www.ssajm.org/text.asp?2014/1/3/111/138930>
- Vichinsky et al. Causes and outcomes of the acute chest syndrome in sickle cell disease. N Engl J Med 2000; 342:1855-1865 DOI: 10.1056/NEJM200006223422502





# Appreciation



A graphic for World Sick Cell Day Awareness Day. The background is dark with various red, 3D-rendered sickle cells and normal red blood cells. A red diagonal band at the bottom contains the text.

WORLD

# SICKLE CELL DAY

AWARENESS DAY - JUNE 19TH



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