SICKLE CELL DISEASE: ADDRESSING RACISM & DISCRIMINATION

Kimberly Sapre, DMSc, PA-C, CAQ-EM

Quality Director for the Emergency Medicine APPs

Emergency Medicine PA

Inova Fairfax Medical Campus, Falls Church, VA

DISCLOSURES

I have no relevant relationships with ineligible companies to disclose within the past 24 months.

OBJECTIVES

At the conclusion of this session, participants should be able to:

1. Recall the pathophysiology and epidemiology of sickle cell disease
2. Recognize the acute nature of sickle cell disease leading to recurrent emergency department visits
3. Identify treatment barriers preventing adequate pain management in the emergency department

TREATMENT BARRIERS

- ★ Knowledge deficit
- ★ Stigma and perception of addiction
- ★ Frequent ED visits
- Lack of primary care clinicians
- Lack of access to specialized care
- ★ Inadequate treatments

Racism & Discrimination

RACISM & DISCRIMINATION

Stereotype

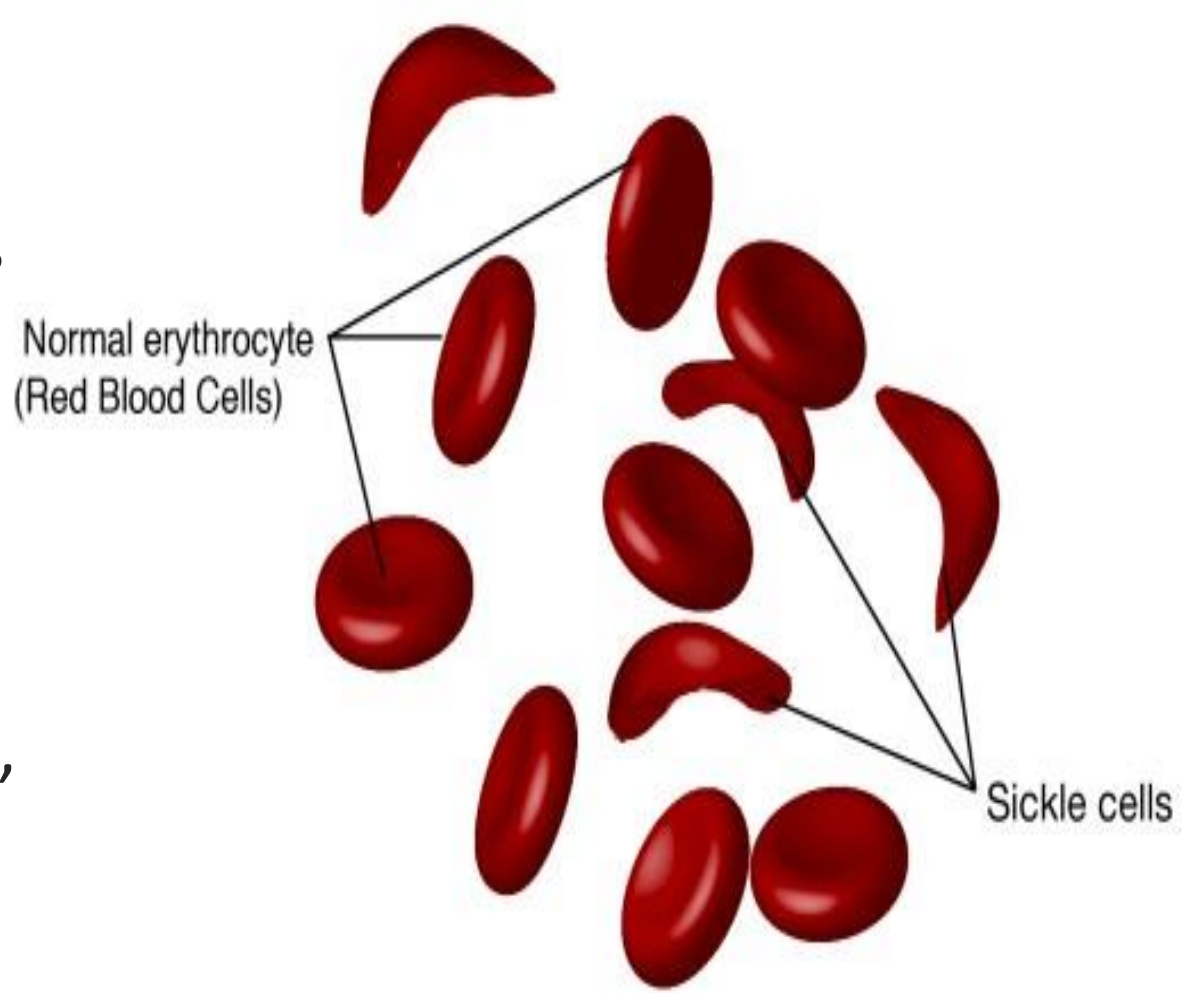
Prejudice

Discrimination

Racism

SICKLE CELL DISEASE

- Most common genetic disorder
- Recessively inherited disorder
- Deoxygenated hemoglobin aggregates into rigid polymers, resulting in “sickle”-shaped erythrocytes
- Average life expectancy: 29 – 54 years old
- Curative options : Stem cell transplant, CRISPR gene editing



- Most common in people of African descent
- Hispanic/Latino, South Asian, Middle Eastern, and Mediterranean descent
- More than 55% of patients reside in 10 states

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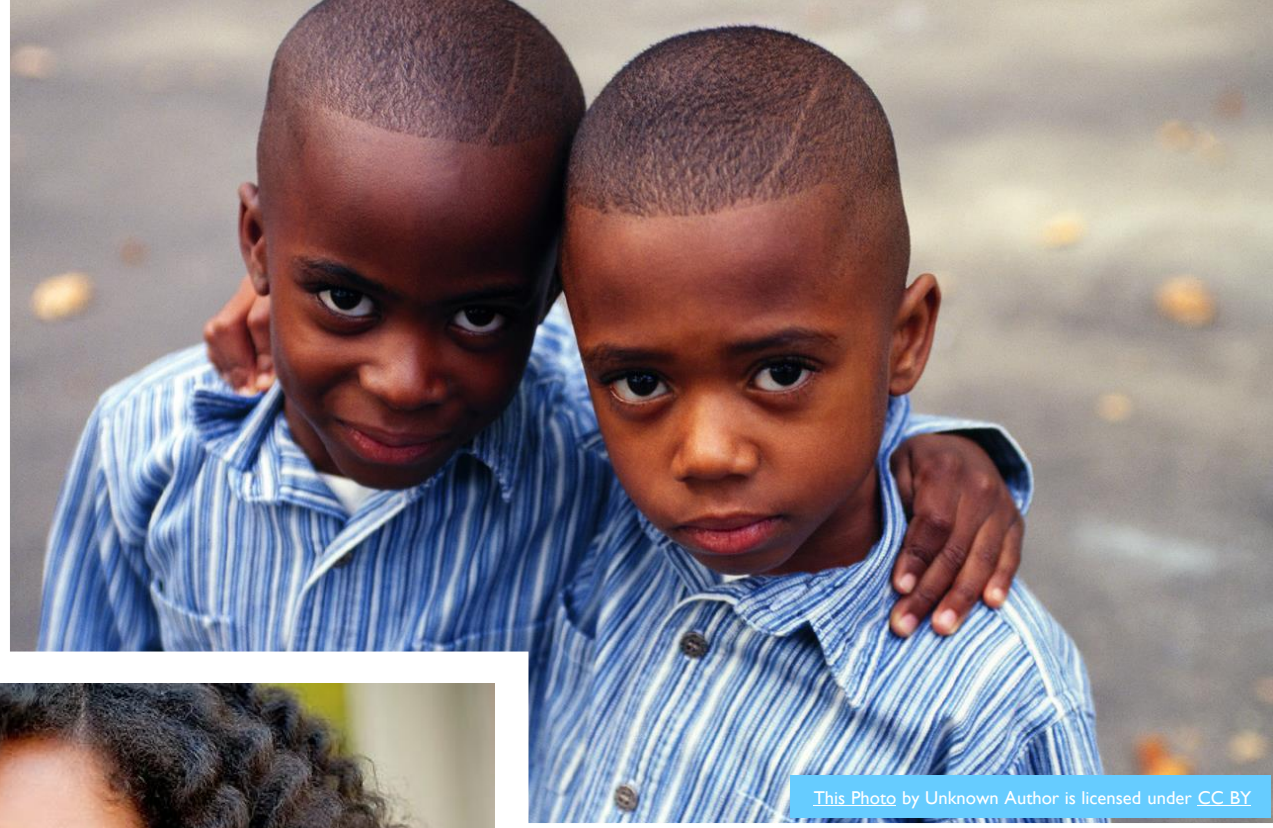

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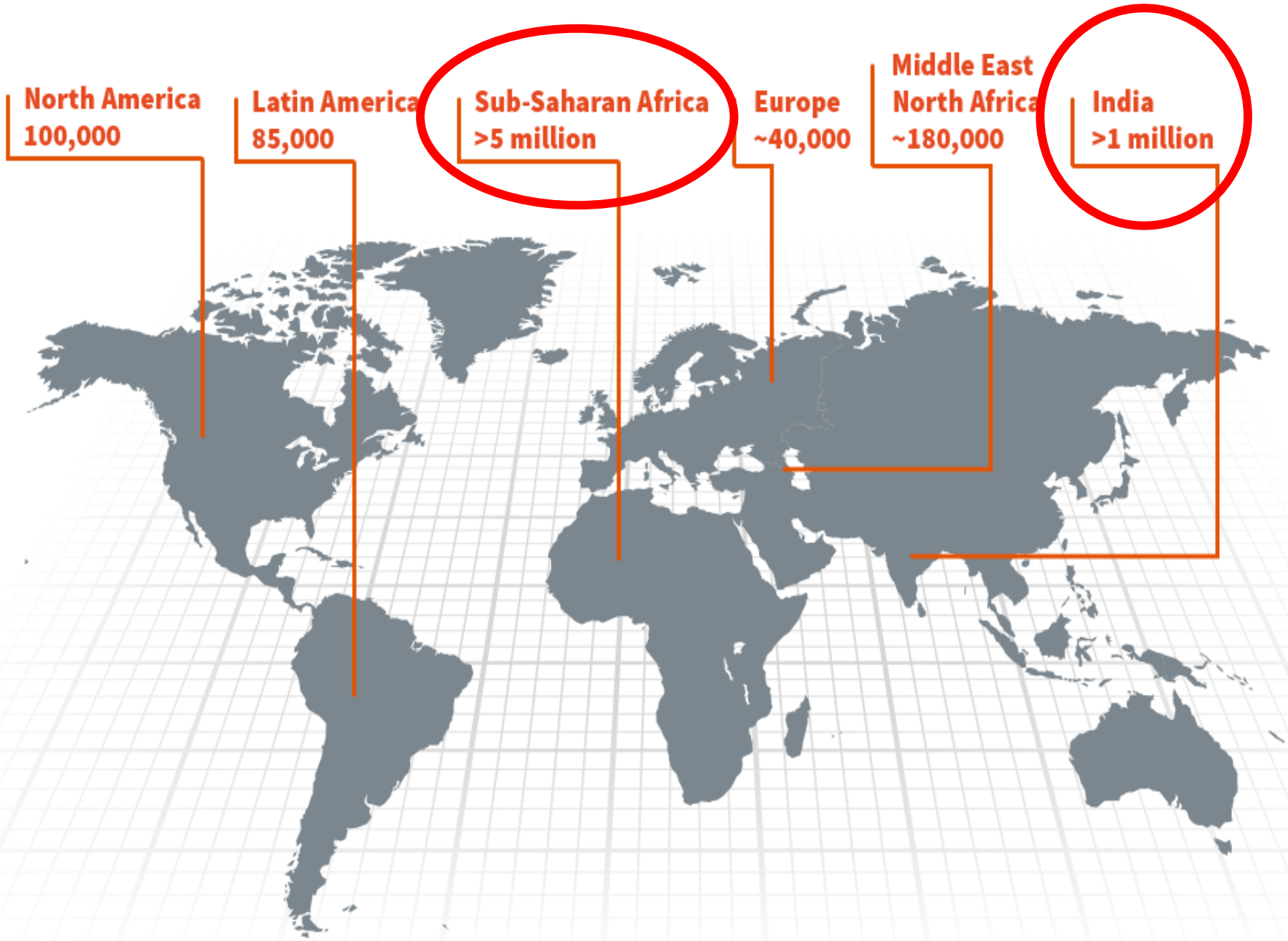
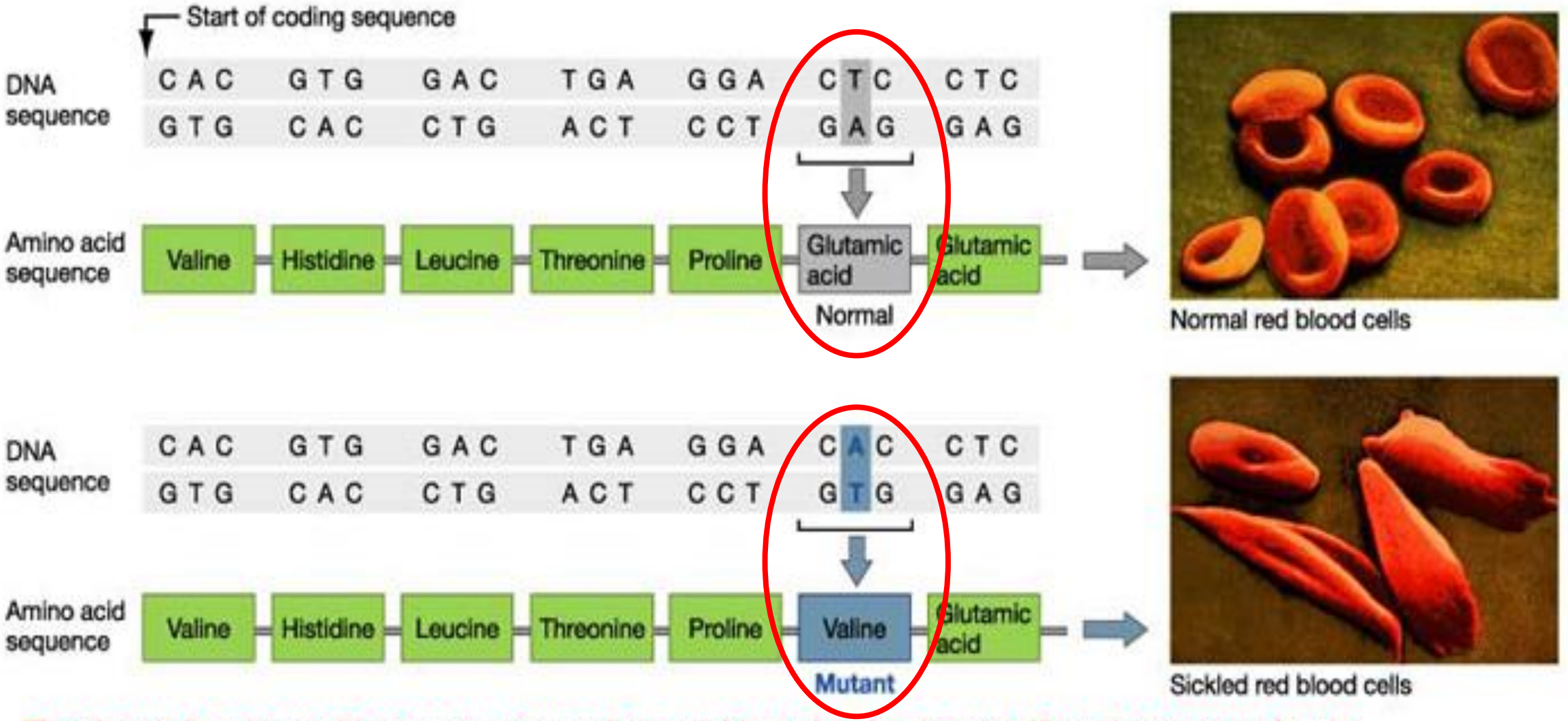
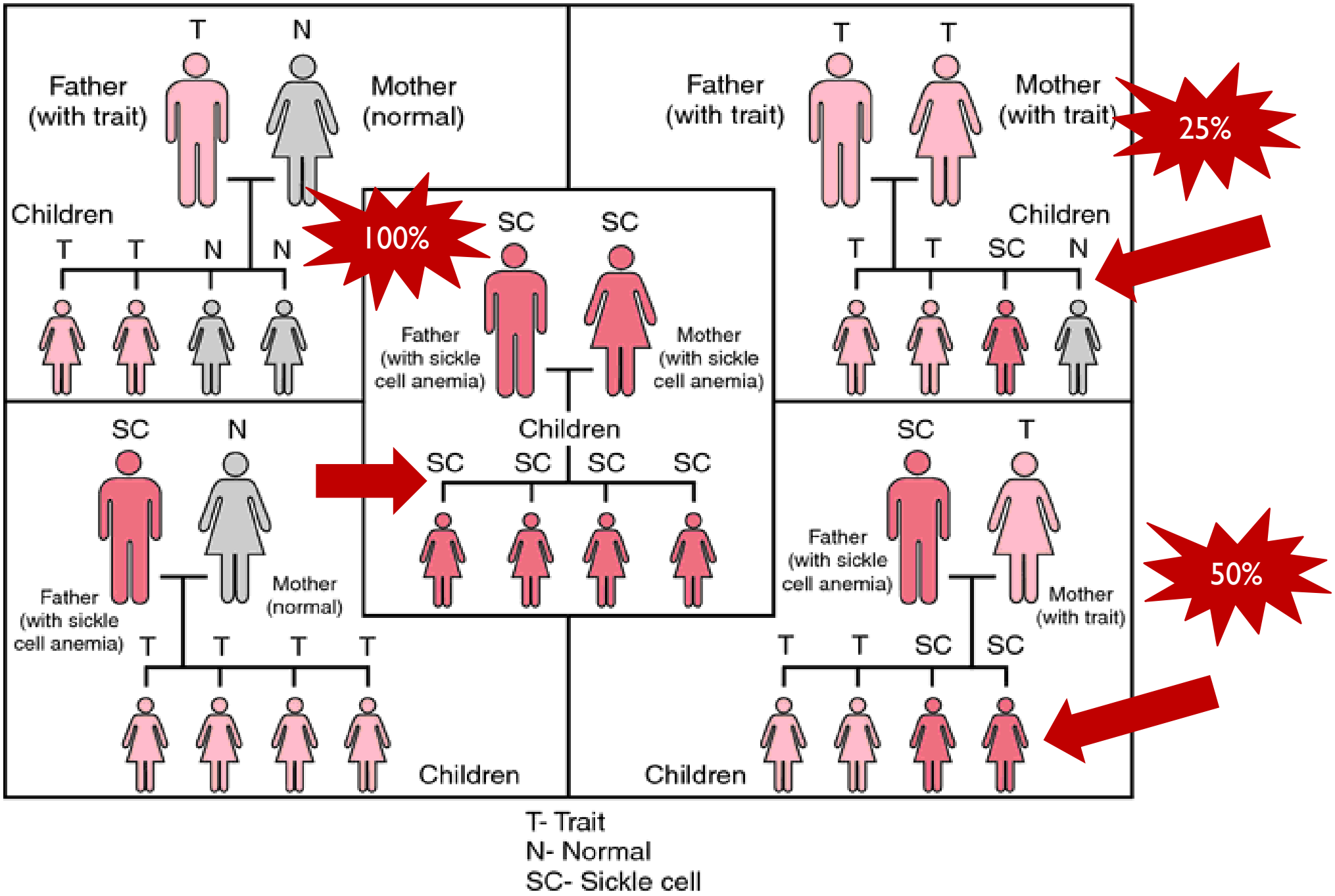

North America
100,000
Latin America
85,000
Sub-Saharan Africa
>5 million
Europe
~40,000
Middle East
North Africa
~180,000
India
>1 million



The change in amino acid sequence causes hemoglobin molecules to crystallize when oxygen levels in the blood are low. As a result, red blood cells sickle and get stuck in small blood vessels.


T
N
Father
(with trait)
Mother
(normal)
Children
T T N N
100%
SC SC
Father
(with sickle
cell anemia)
Mother
(with sickle
cell anemia)
Children
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T T SC SC
Children
T- Trait
N- Normal
SC- Sickle cell

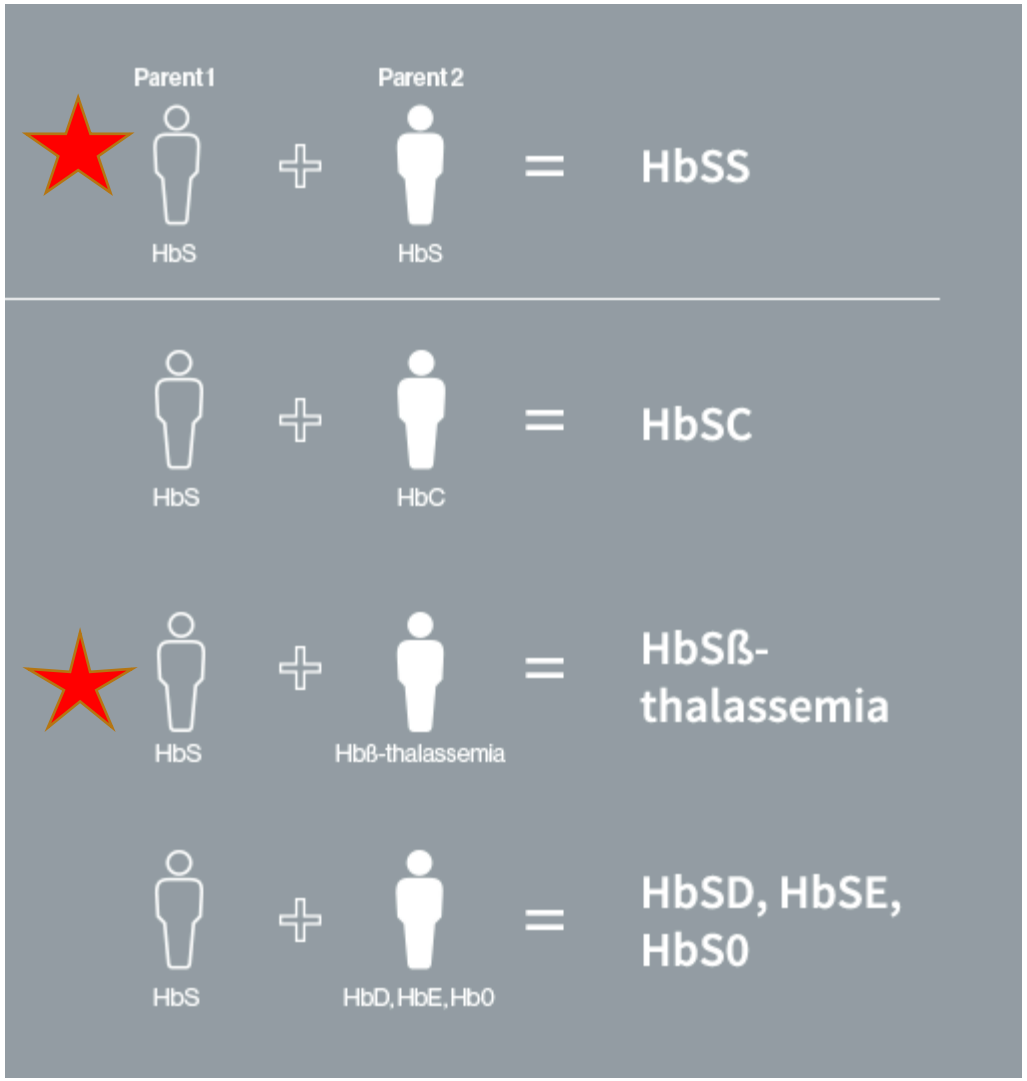
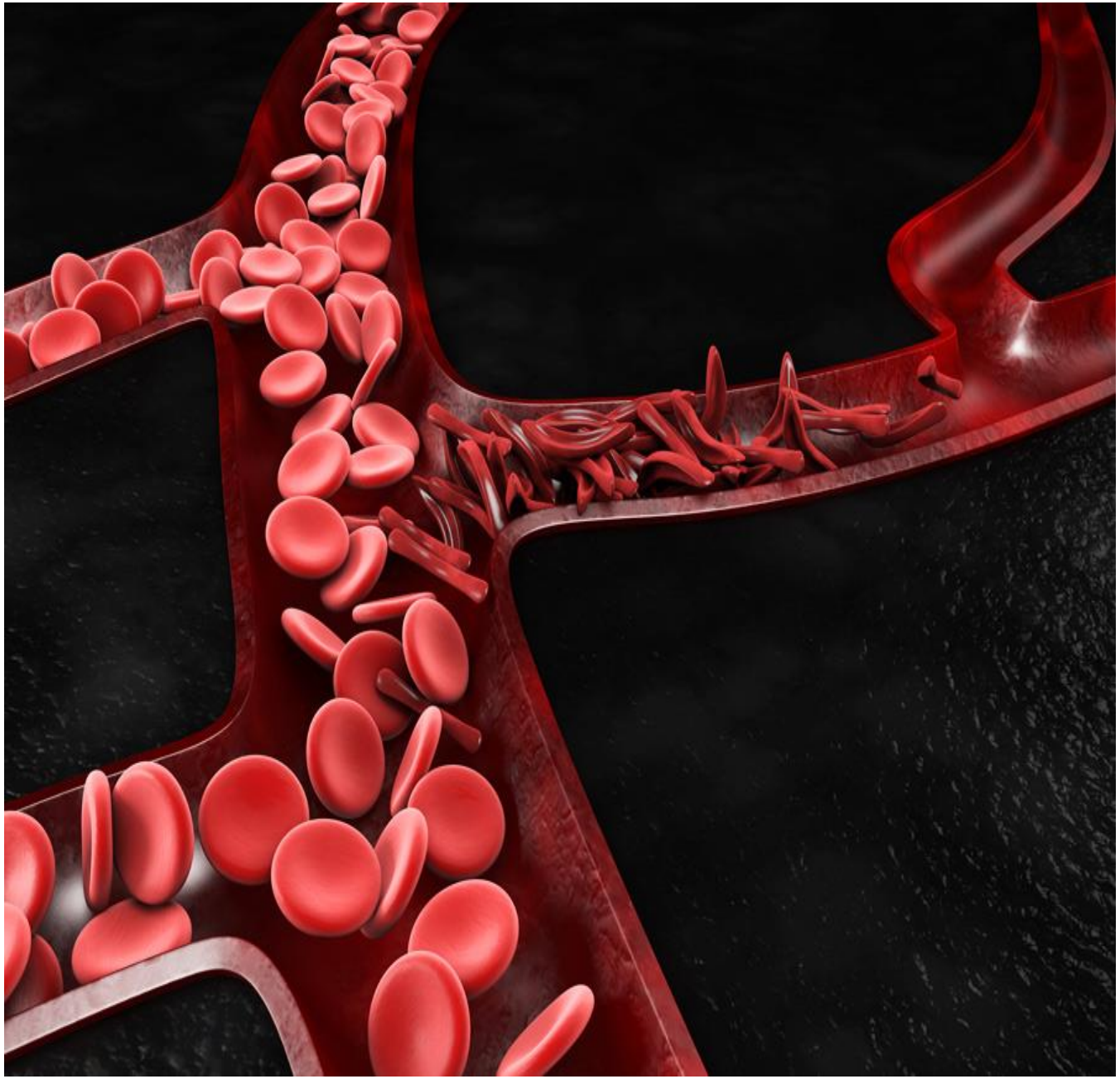


Table 1
Genotypes and phenotypes of different sickling disorders

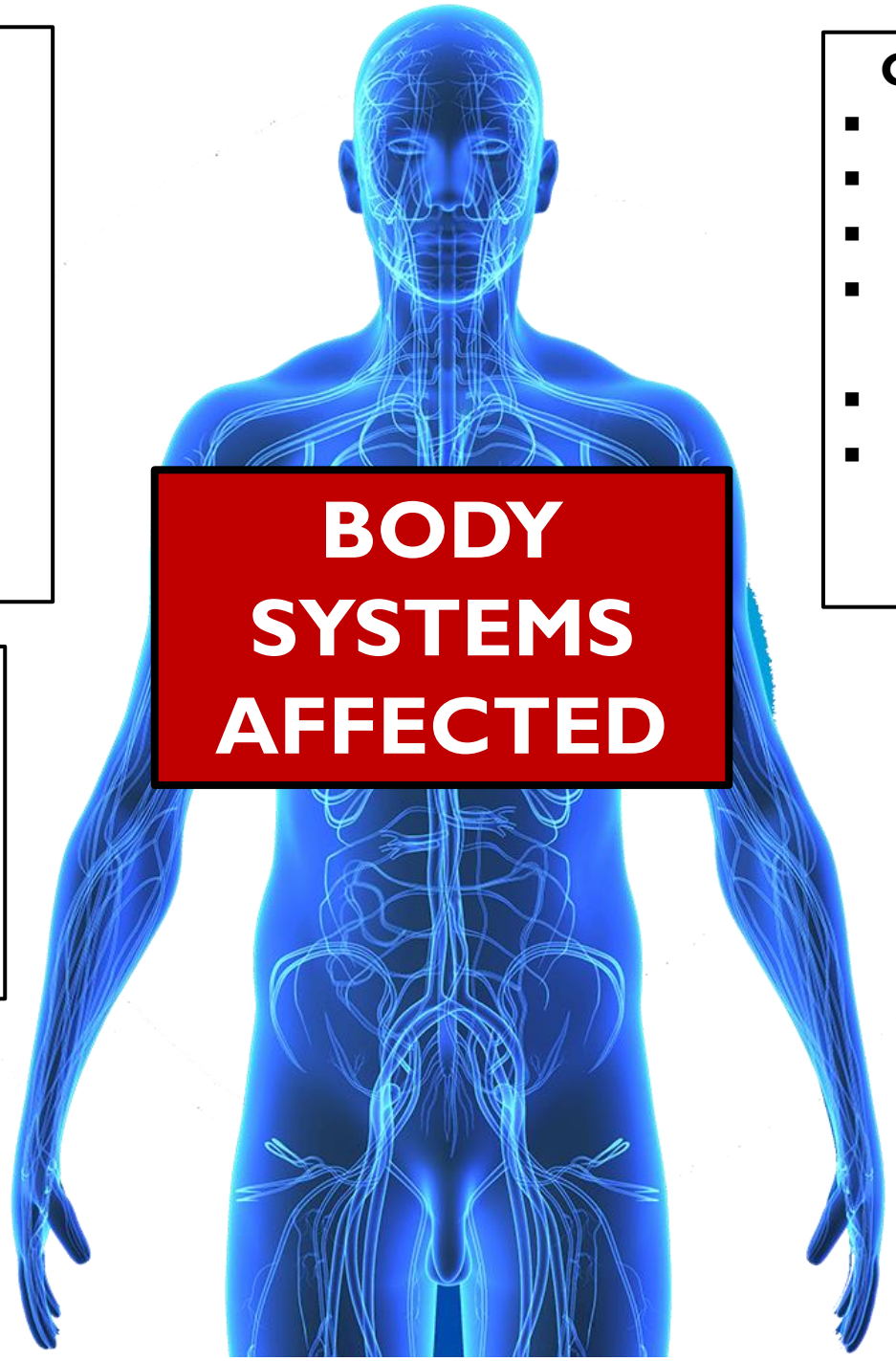
Genotype	% of Hb Type/Total Hb in a Typical Patient					Clinical Course
	HbS	HbA	HbF	HbC	HbA2	
Not SCD						
HbAA (normal)	—	96	2	—	2	No manifestations
HbAS (trait)	45	50	2	—	2	No manifestations[a]
SCD						
HbSS	95	—	3	—	2	Severe
HbSC	48	—	3	47	2	Moderate
HbS β^0	93	—	2	—	5	Severe
HbSβ^+ (moderate)	85	6	5	—	4	Moderate
HbS-β^+ (mild)	70	23	3	—	4	Mild

VASO-OCCLUSIVE CRISIS

- Sickled cells →microvascular occlusion → ischemia and hypoxia → pain
- Pediatric patients may have their first crisis within the first year of life
- 50% of HbSS patients will have the first crisis by 5 years old



BODY SYSTEMS AFFECTED



NERVOUS

- Acute ischemic stroke
- Hemorrhagic stroke
- Venous sinus thrombosis
- Proliferative retinopathy
- Silent infarction
- Orbital infarction
- Chronic pain

Cognitive Impairment

CARDIOPULMONARY

- Acute chest syndrome
- Pulmonary embolism
- Pulmonary hypertension
- Chronic restrictive lung disease
- Dysrhythmias
- Deep Venous Thrombosis

Sudden death

RETICULOENDOTHELIAL

- Splenic sequestration
- Functional hyposlpenism

Anemia

Hemolysis

GASTROINTESTINAL

- Cholelithiasis/cholecystitis
- Hepatopathy
- Mesenteric vaso-occlusion

UROGENITAL

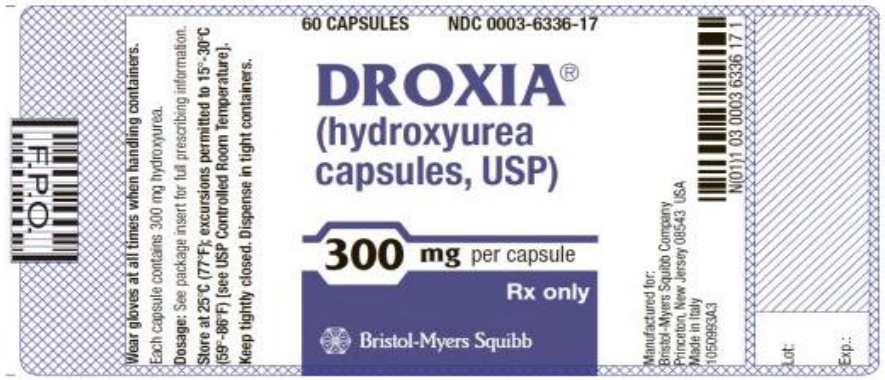
- Papillary necrosis
- Proteinuria
- Renal Failure
- Hematuria
- Nocturnal enuresis
- Priapism

MUSCULOSKELETAL

- Avascular necrosis
- Leg ulceration
- Osteomyelitis

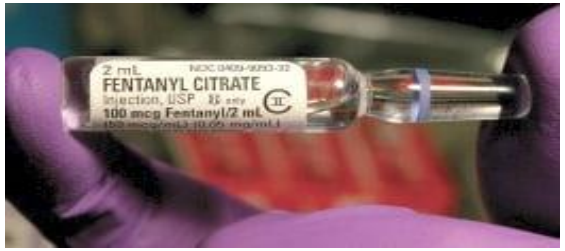
INADEQUATE TREATMENTS

TREATMENTS



voxelotor

crizanlizumab



RESEARCH DISPARITIES

SICKLE CELL DISEASE

- 1 in 365 Black individuals
- NIH Funding per person $807
- Annual PubMed Publications 926
- New FDA drug approvals 1
- Novel FDA drug indications 2

CYSTIC FIBROSIS

- 1 in 2500 white individuals
- NIH Funding per person $2807
- Annual PubMed Publications 1594
- New FDA drug approvals 4
- Novel FDA drug indications 11

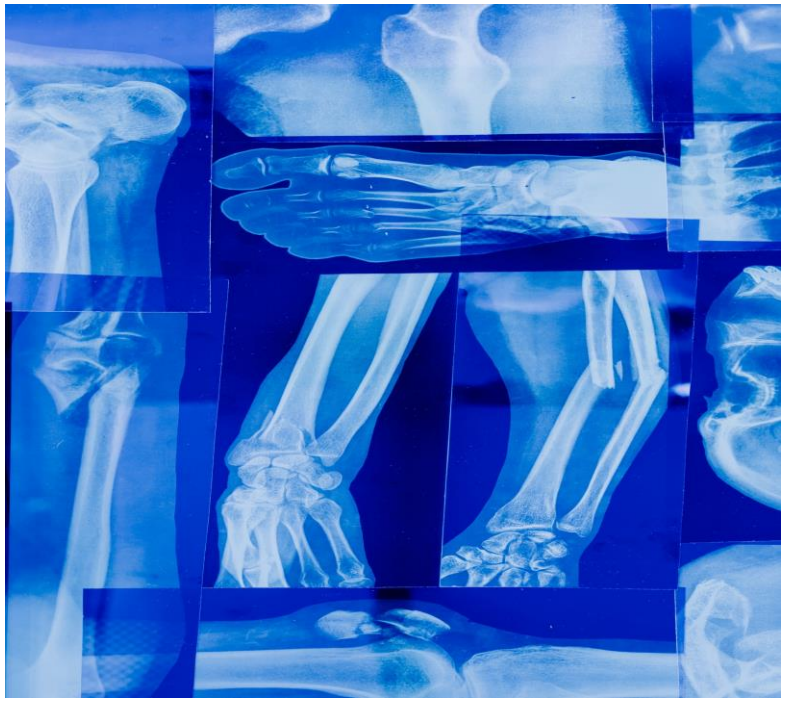
OPIOID CRISIS

OPIOID ABUSE

- 2008-2013 opioid use stable in SCD population
- Less than 1% of drug overdose deaths
- Requesting a specific opioid is NOT drug seeking behavior
- Trust the patient's assessment of pain
- Do not refer to patients as "sicklers"

EMERGENCY DEPARTMENT

- More likely to present with severe pain (7-10)
- 25% longer wait times than general sample
- SCD patients waited 50% longer than long bone fracture patients for pain medications
- SCD wait times similar to other Black patients with other complaints



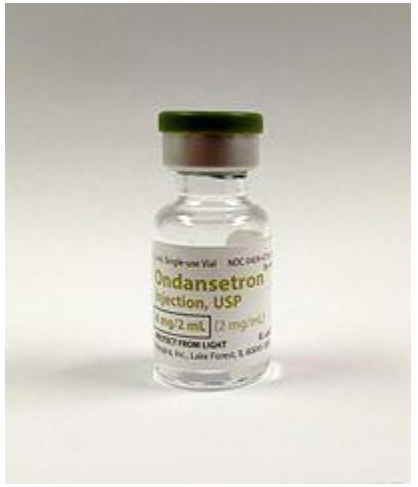
WHAT CAN WE DO?

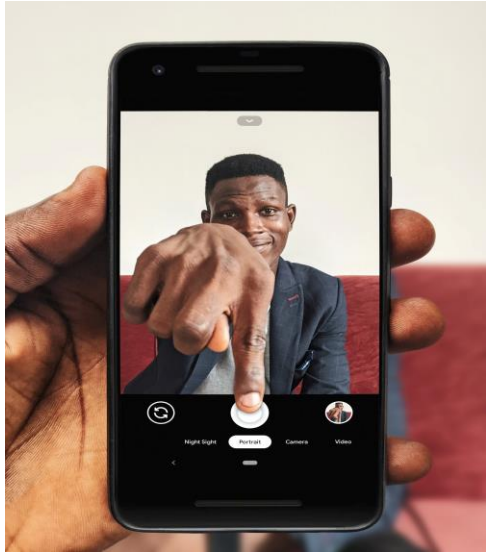
ED TREATMENT PROTOCOLS

- All patients triaged to Level 2
- Trust the patient's assessment of their pain level
- Ask the patient for the medications that have worked in the past
- Give pain medications within 60 minutes of arrival
- Give opiate medications every 15-30 minutes, reassessing pain
- If pain not well-controlled, admit for pain control

ED TREATMENT PROTOCOLS

- Treatment adjuncts
- Opioid adverse affects
- Nonpharmacologic interventions - distraction techniques, warm packs, blankets
- Avoid – steroids





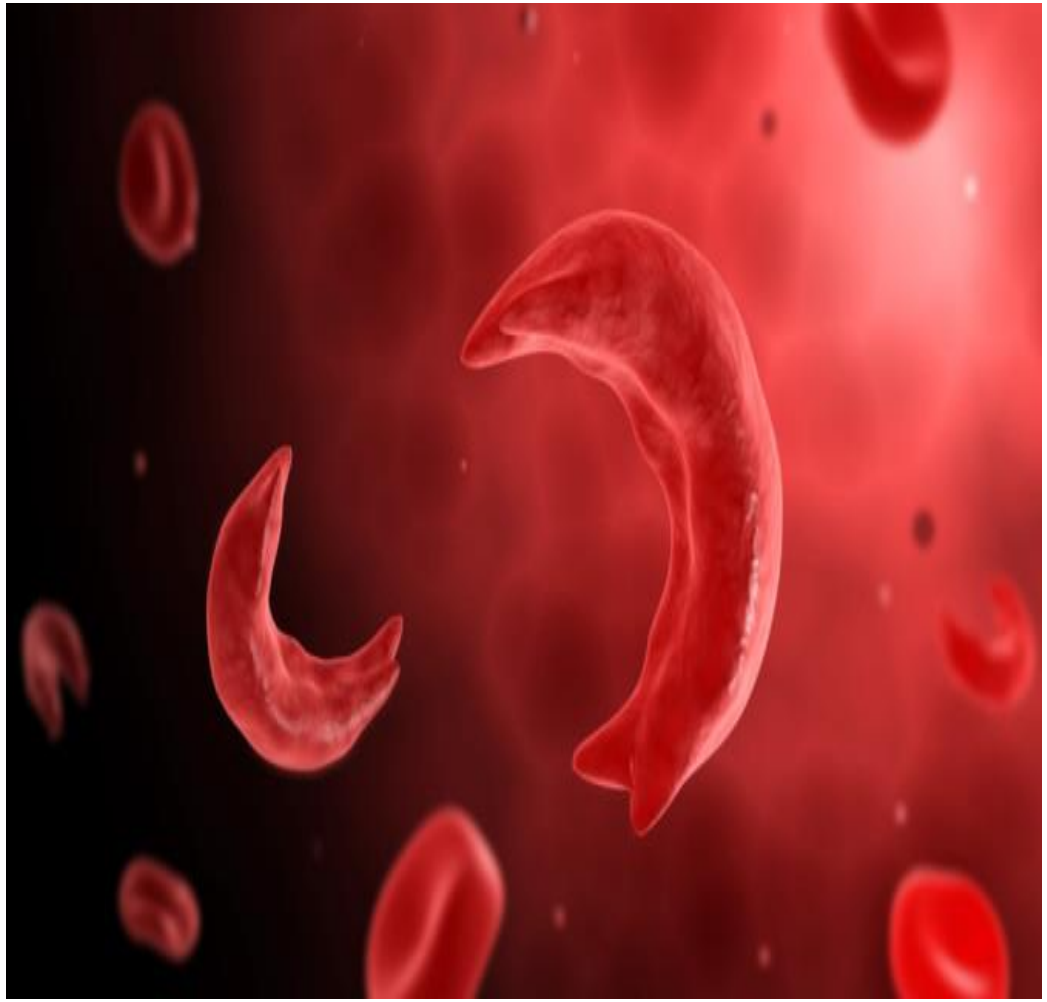
CLINICAL TRIALS
U.S. NATIONAL LIBRARY OF MEDICINE

- Patients and families may register for clinical trials
- Clinicians can learn about the latest trials completed

clinicaltrials.gov

CONCLUSION

- SCD is a genetic disorder which causes severe pain
- Trust patient's description of pain
- Opioids are required for treatment of acute sickle cell crisis
- Do not withhold medications awaiting laboratory results
- Develop a departmental protocol to quickly treat pain



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QUESTIONS??

kimsapre@gmail.com
@KimEMPA2019