Resilient Bodies and Voices Unheard: A Sickle Cell Disease Case Study

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“Hope” and “Need not suffer alone”
- Hertz Nazaire

Clinical Pearls from the Experts: Dr. Vichinski, Dr. Hagar, and Dr. Thompson

Eliott Vichinski

Pediatric hematologist-oncologist
Director, Thalassemia Center
Director, Pediatric Sickle Cell Anemia Center

Alexis Thompson

Pediatric hematologist-oncologist
Chief of the Division of Hematology at Children's Hospital of Philadelphia
Former ASH President (2018)

Robet Ward Hagar

Internist
Director of Adult Sickle Cell Center at UCSF Benioff Children's Hospital Oakland.

A few quick questions to test the water:

Which of the following are reliable, objective indicators of acute sickle cell disease (SCD) pain

a. Elevated blood pressure
b. Blood/Oxygen saturation level below 92%
c. Hemoglobin below 8
d. Retic Count > 15%
e. All of the above
f. None of the above

A few quick questions to test the water:

F. None of the above

Rationale: There are no reliable, objective indicators of acute SCD pain.

Patient self-report is the **gold standard** of pain measurement

One of the biggest misunderstandings is looking at hemoglobin as a guide to a person's pain. Pain can be from hemolysis, hyperviscosity, allodynia, neuro-inflammation…all distinct from hemoglobin levels.

Yang F, Smart Health 2019

A few quick questions to test the water:

- How long does a vaso-occlusive crisis typically last?

A few quick questions to test the water:

Length of time varies, but this can be summarized into four acute phases:

- Chronic: Patients are in a chronic state of pain.
- Phase 1: The prodromal phase, lasting 1-2 days, patients often describe ‘achy-ness’ and numbness/paresthesias.
- Phase 2: Maximal pain is reached rapidly due to local tissue infarct from vaso-occlusion. A transition from achy to stabbing pain.
- Phase 3: Post-infarct inflammatory responses lead to pain becoming constant and often associated with a fever, lasting 3 to 5 days.
- Phase 4: Resolution of symptoms, often lasting 1-2 days.

Jang T, J Transl Med. 2021

A few quick questions to test the water:

What is the average life expectancy for a person living with Sickle Cell Disease in:

- The United Kingdom ___
- The United States ___
- Nigeria ___
- California ___

A. 21
B. 52.6
C. 39
D. Mid-60's (Median of 67)

A few quick questions to test the water:

The average life expectancy for a person living with Sickle Cell Disease in:

United Kingdom	D. Mid 60’s, median age of 67
United States	B. 52.6
Nigeria	A. 21
California	C. 39

Case Presentation: Marcus - 35M

35M w/hx of SCD (HbSS) presenting w/ acute onset left hip pain

- For some more context, here are some facts that were found in different areas of the EMR:
  - Patient has had 15 ED visits in the 6 months for pain crises
  - He was labeled as “drug seeking” and “frequent flyer”
  - Also described as opioid dependent and tolerant

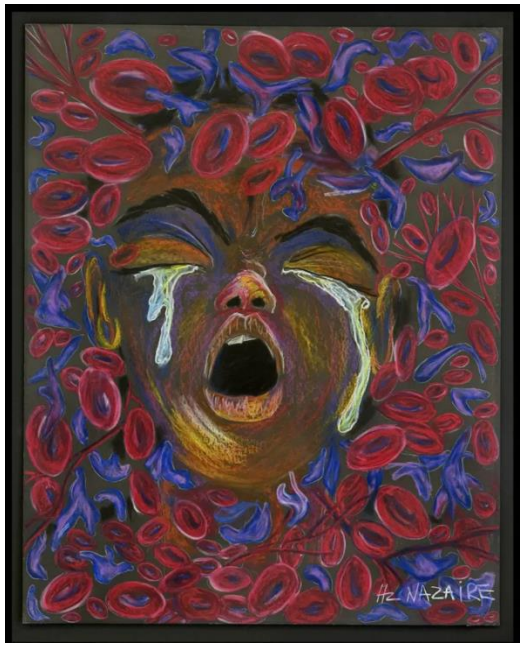
Jan	Feb	Mar	Apr	May	Jun
1/4	2/1	3/4	4/10	5/4	6/4
1/18	2/20		4/14	5/12	6/12
	2/29		4/20	5/16	
			4/28	5/24	

Case Presentation: Marcus - 35M

Triage Note: Pt states “Pain began a couple days ago, I am having a Sickle Cell crisis. 10/10 pain.” He added “I can’t even hold food, fluid, and my pain medications.” Here 2 days ago with similar pain crisis.

Triage Note: R sided abdominal pain since Wed, intermittent fever, HR 146

Triage Note: Generalized weakness x 1 week, BIBA from SNF



“10 Redefined”
- Hertz Nazaire

TT	Pt Rm Time	Bed	Patient	Age	ESI	Temp	BP	HR	SpO2	RR
4:06	00:06	Room 22	Marcus (M)	35	3	37.9	142/87	91	96%	20

Case Presentation: Marcus - 35M

Marcus is a 35M w/hx of sickle cell disease (Hgb SS), avascular necrosis of left hip in setting of recurrent osteomyelitis, asplenia, who was seen 8 days ago for pain crisis, presenting w/right leg pain. Recently seen on 6/4 for acute pain crisis, admitted for crisis management and discharged on 6/7. Patient is wheelchair bound since March due to avascular necrosis of the left hip. Over the past 5 days, patient has had worsening, generalized pain in his upper legs, particularly in the right leg. **This pain is different from the pain he typically has**, it is usually the left leg that is painful. He describes the pain as 10/10, sharp and has not had any relief from his at home medications. **He has been unable to sleep for the last few days due to the pain**. He describes feeling more dehydrated due to the heat outside and having to use his wheelchair to get around. Patient has also been having **new left leg numbness**, that has been there for a few months, no other FND. He denies any fever, chills, no CP/SOB, abdominal pain, rhinorrhea/congestion, urinary or stool concerns, and no recent sick contacts. When having a crisis, Benadryl 25 mg, Dilaudid 2 mg, Zofran 4 mg, Toradol 30 mg, and IV fluids work best.

TT	Pt Rm Time	Bed	Patient	Age	ESI	Temp	BP	HR	SpO2	RR
4:06	00:06	Room 22	Marcus (M)	35	3	37.9	142/87	91	96%	20

Clinical Pearls from the Experts: Dr. Vichinski, Dr. Hagar, and Dr. Thompson

- A key question to always ask: is this typical for your pain?
  - For many patients, vaso-occlusive crises often follow similar patterns.
  - When patients describe that this is a new type of pain or something different, it should raise concern.
- An important question to never ask: how long have you had Sickle Cell Disease?
  - You may be surprised how often this is asked.
- Always ask a patient whether they have a pain plan and if not what treatment works best for them.
  - You may be surprised at the recommendations you receive from a patient, they are not always what you would expect.
- Sleep, or lack thereof, is not something always considered in a patient's presentation.
  - It is important to understand that the immense pain hinders sleep and most often patient's are presenting days into a crisis. Once pain is under control, they can fall asleep.

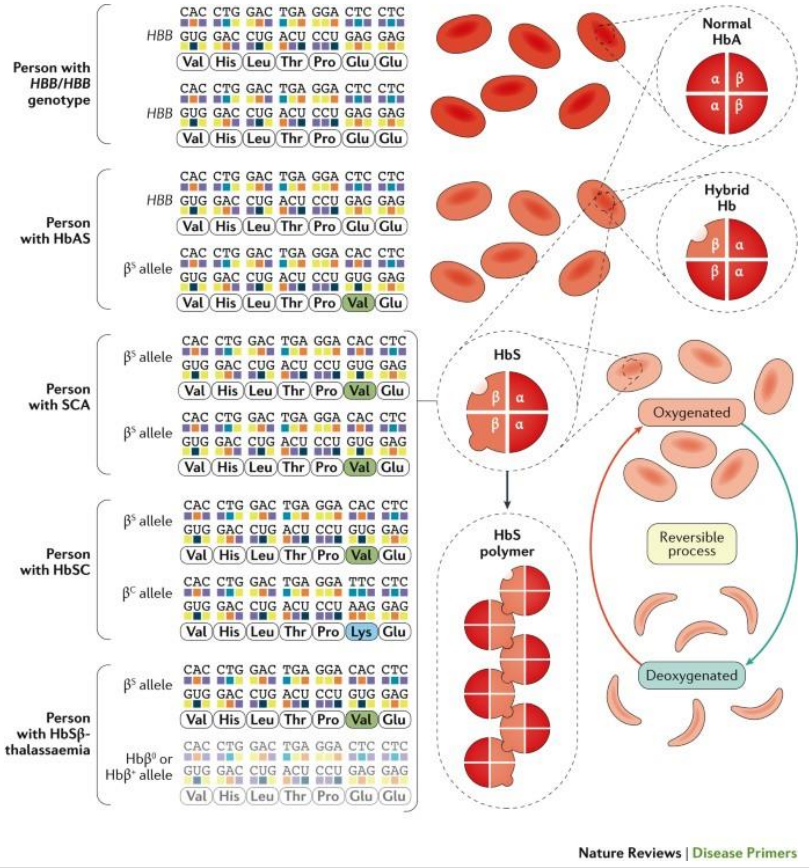
Sickle Cell Disease: What is it?

Sickle Cell Disease: a group of inherited diseases including Sickle Cell Anemia (SCA), HbSC, and HbSβ

- Hemoglobin is a tetrameric protein composed of different globin subunits, each of which has the cofactor heme, that can carry a molecule of oxygen
- A single nucleotide substitution in the sickle β-globin unit, leads to an amino acid substitution (Glutamic Acid → Valine)
- Under conditions of deoxygenation (Hb not bound to oxygen), the mutant sickle β-globin units can polymerize and cause the erythrocytes to assume a crescent or sickled shape

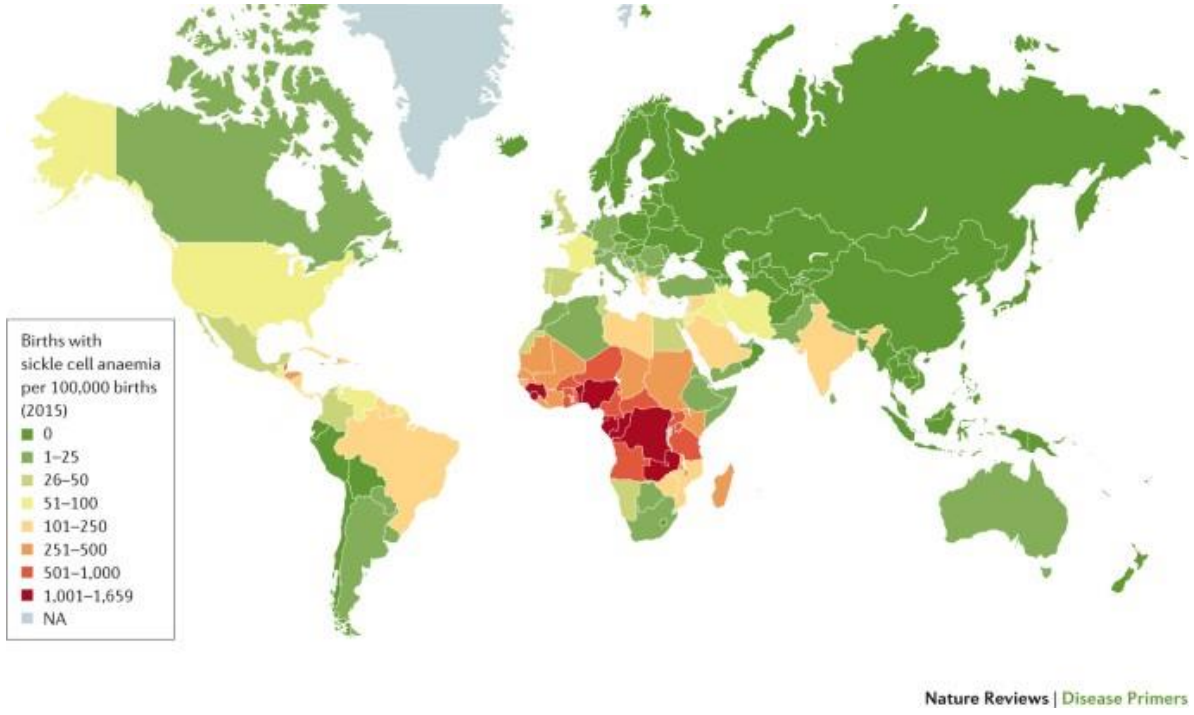
*** It is important to understand that the transition from hemoglobin F to hemoglobin SS occurs by 9 months

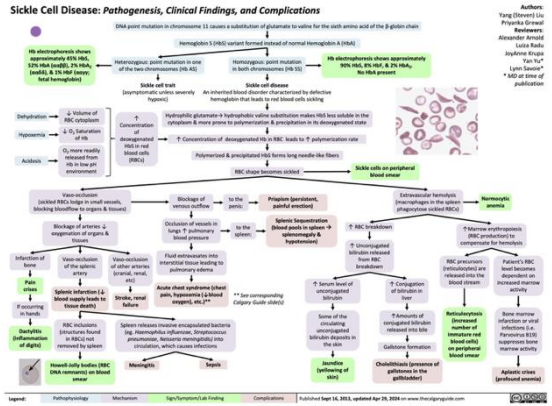
- That is right, sickling (pain) already occurs in 9-month-old infants

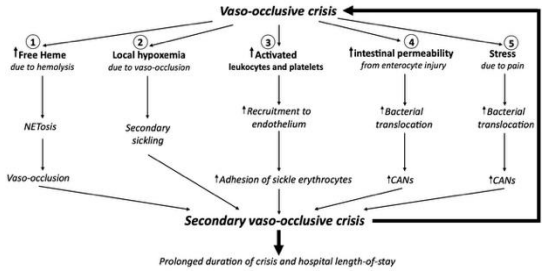


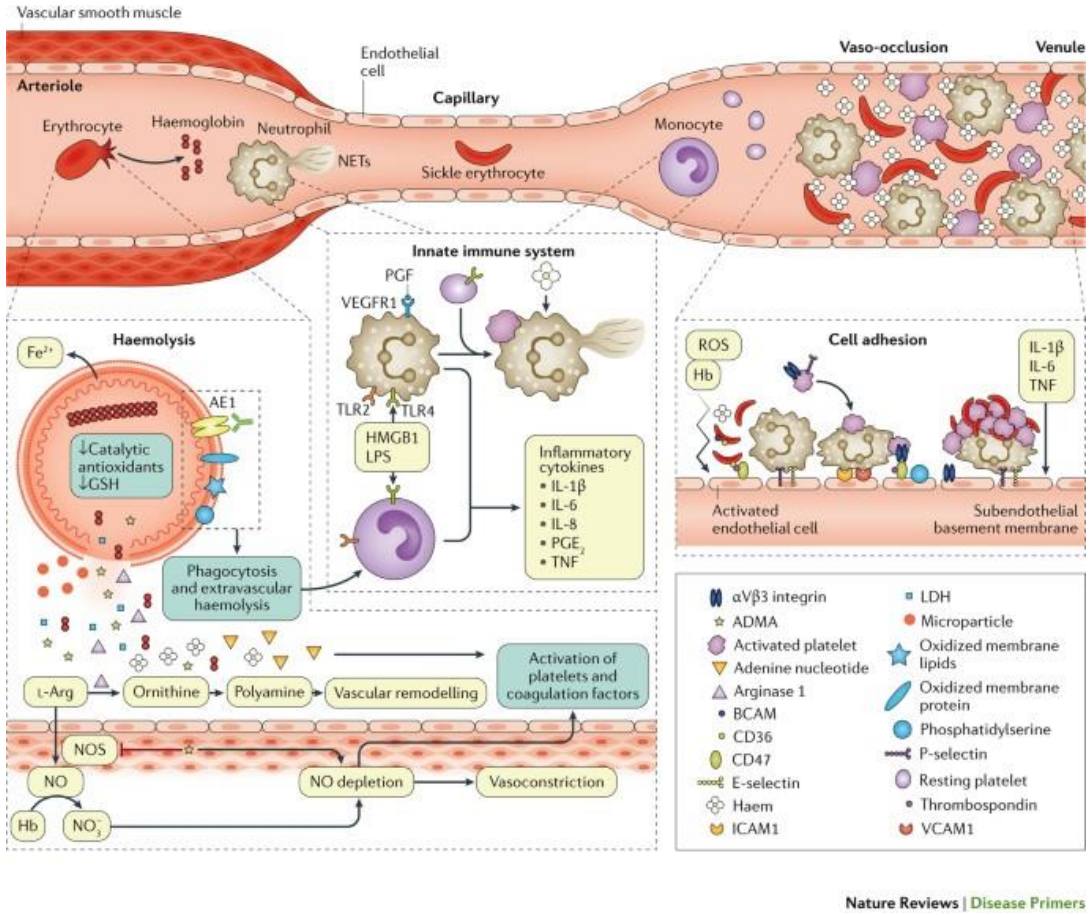
Epidemiology

- Geographical Distribution is driven by:
  - The endemicity of malaria and population movements
  - Those with sickle cell trait (HbAS) have remarkable protection against severe P. falciparum malaria (90% less likely to experience severe malaria than those with only normal Hb)
  - This explains the high frequency of the β^S sub-Saharan Africa, the Middle East, India, and the Mediterranean.
  - Population movements, including the slave trade led to a much wider distribution, particularly in North America and Western Europe









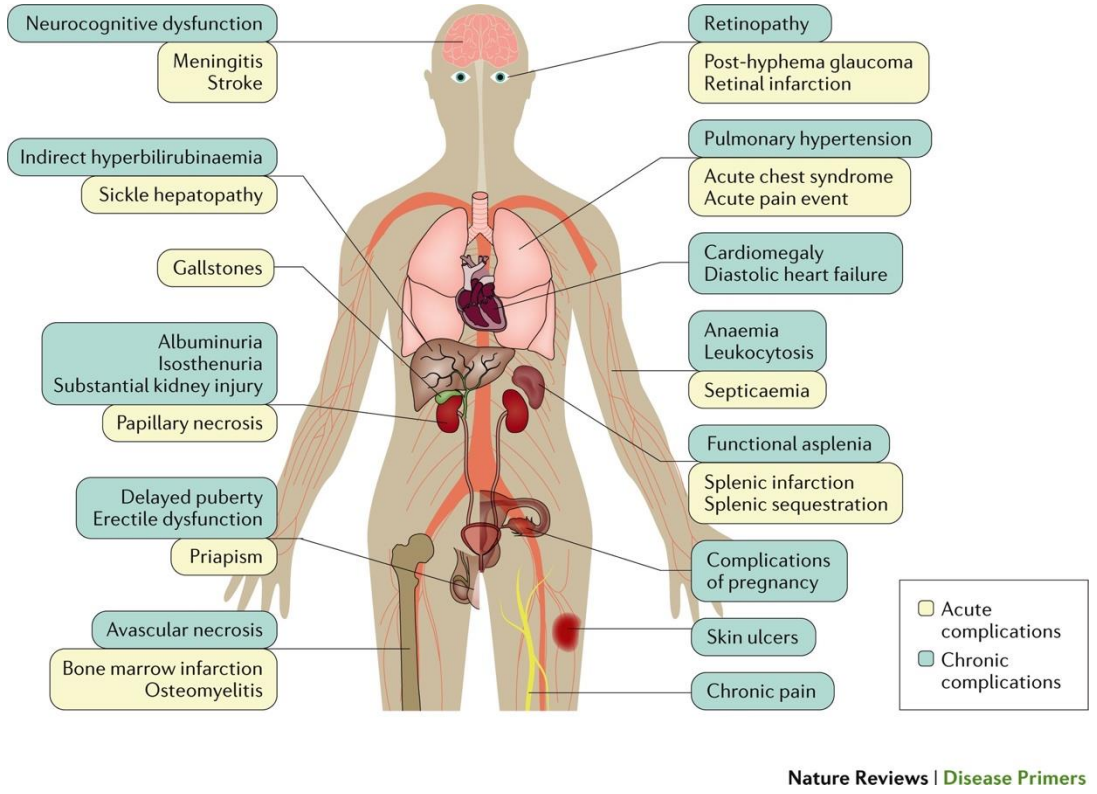
Pathophysiology

- Sickled cells can represent anywhere from 5 to 50% of circulating erythrocytes, resulting in chronic, ongoing hemolysis and episodic periods of vascular occlusion.
- Damage of the RBC membrane caused by β^S leads to both extravascular and intravascular hemolysis.
- The contents of the RBCs lead to a whole lotta problems:
  - an increase in reactive oxygen species
  - activation of platelet and blood clotting proteins
  - activation of the innate immune system (presence of Heme)
  - release of inflammatory cytokines from activated monocytes and macrophages which activate the endothelium (sticky)
  - formation of Neutrophil Extracellular Traps which bind activated platelets and sickle cell erythrocytes forming aggregates (causing vaso-occlusion)

Clinical Complications

SCD is a complex, multisystem condition characterized by acute and chronic complications.

We will home in on a few: bones, chest, blood, and brain.



Case Presentation: Marcus - 35M

Marcus is a neighborhood, sickle cell patient with chronic left hip osteomyelitis who comes to the ED stating he has 10/10 pain "all up in my arms and legs." He is narcotic dependent, and here frequently. At home he reportedly takes 100 mg OxyContin BID and oxycodone 5 mg for breakthrough pain. Over the past few days, he says that he has taken 2 tabs every 4–6 hours. About 3 months ago, patient states that after getting out of the SNF, the housing authority moved him to a new neighborhood, and he now has to wheel himself in a manual wheelchair up 3 blocks from the bus stop. Yesterday afternoon, he was hanging out with friends outside McDonald's where he wheeled himself around more than usual and got dehydrated due to the heat. He believes that this, along with some "stressful situations," has precipitated his current crisis. Pain is sharp in quality, right sided, severe (10/10), and has not been helped by any of the narcotic medications he says he has already taken. On physical exam, he appears to be in distress. He has no fever, and his pulse ox is 96% on RA. The rest of the physical exam is normal although he reports decreased sensation in the left hip/leg.

TT	Pt Rm Time	Bed	Patient	Age	ESI	Temp	BP	HR	SpO2	RR
4:06	00:06	Room 22	Marcus (M)	35	3	37.9	142/87	91	96%	20

What is a “sickler”? Do words matter?

The Impact of Heuristics

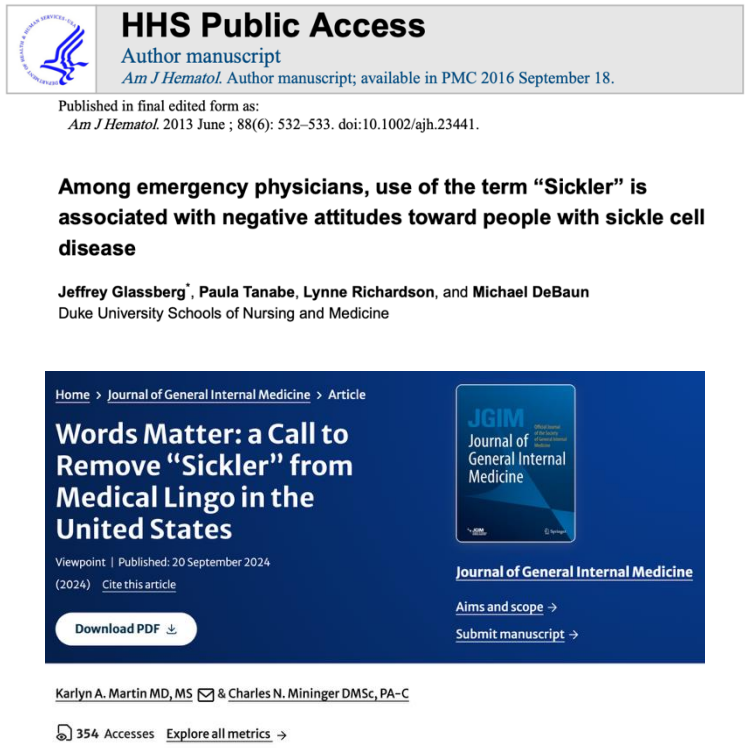
- Heuristics in medicine: mental shortcuts that enable rapid decision making, simplify complex problems into manageable components, promote consistency in decision making, and reduce cognitive load.
- Conversely, however, heuristics can reinforce biases and stereotypes, and lead to an oversimplification of presenting information and results in cognitive distortion.

Words Matter: a Call to Remove “Sickler” from Medical Lingo in the United States

- ”Sickler” has undergone a connotative shift, it is now one increasingly associated with stereotypes related to health, race, and socioeconomic status.
    - The disease is strongly associated with race (> 90% off patients are Black American), carrying with it the unconscious bias of structural and institutional racism that already exist.
    - An association with pain. Pain is a real consequence of ischemia and is a symptom during a time at which patients are at high risk for poor outcomes.

Among emergency physicians, use of the term “Sickler” is associated with negative attitudes toward people with sickle cell disease

- 655 emergency physicians from 49 states with representation from academic (67.9%) an community (32.1%) providers
- A statistically significant relationship was observed between physician negative attitudes toward individuals with SCD and the use of the term “sickler.”



HHS Public Access

Author manuscript

Am J Hematol. Author manuscript; available in PMC 2016 September 18.

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Am J Hematol. 2013 June ; 88(6): 532–533. doi:10.1002/ajh.23441.

Among emergency physicians, use of the term “Sickler” is associated with negative attitudes toward people with sickle cell disease

Jeffrey Glassberg[*], **Paula Tanabe**, **Lynne Richardson**, and **Michael DeBaun**

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Words Matter: a Call to Remove “Sickler” from Medical Lingo in the United States

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Martin K, JGIM 2024

Glassberg J, AM J Hematology 2013

Case Presentation: Marcus - 35M

Medications: Atorvastatin, Valsartan, Oxycontin 100 mg BID, and Oxycodone 5 mg q4-6 hrs PRN for breakthrough pain.

Allergies: NKDA

PMHx: SCD (Hgb SS), HTN, HLD, and MDD

PSHx: Splenectomy and cholecystectomy

Social Hx: hx of PSUD, currently in transitional housing since discharged requiring wheelchair from SNF in March

Family Hx: unknown, grew up in foster care

TT	Pt Rm Time	Bed	Patient	Age	ESI	Temp	BP	HR	SpO2	RR
4:07	00:06	Room 22	Marcus (M)	35	3	37.9	142/87	91	96%	20

Clinical Pearls from the Experts: Dr. Vichinski, Dr. Hagar, and Dr. Thompson

- How early in life should a patient with SCD be started on Hydroxyurea?
  - The guidelines from the American Society of Hematology state that children should be started on Hydroxyurea as early as 9 months old.
  - The emergency department can be the only place patients with SCD receive care, be an hydroxyur-advocate, and ask.
- The majority of people with SCD are functionally immunocompromised, functional asplenia often develops within the first year of life.
  - They are particularly sensitive to encapsulated organisms like: Streptococcus pneumoniae, Haemophilus influenzae, and Neisseria meningitidis.
  - Ceftriaxone and Vancomycin, what a beautiful combination unless your allergic (confirm with pharmacist :D)

DeBaun MR, Blood Adv. 2020
Ochocinski D, Front Pediatr. 2020
Giroir B.P., AEM 2020

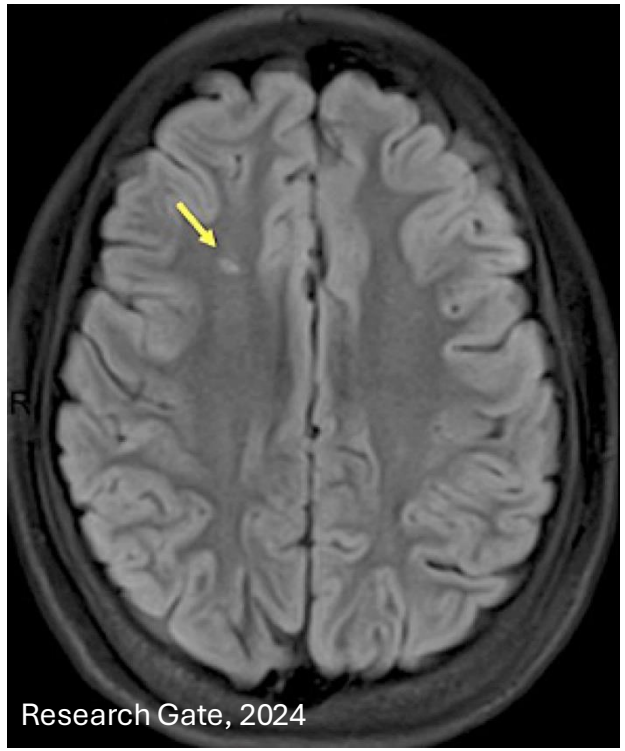
Case Presentation: Marcus - 35M

Physical Exam:			
Gen:	Laying in bed, comfortable in appearance, using his cell phone	Abd:	Minimal tenderness, non-distended
HEENT:	PERRL, EOMI, Oropharynx and TM clear	GU:	No priapism present (up to 30% of males)
Neck:	Trachea midline, no LAD appreciated	Ext:	Point tenderness to palpation in right thigh, otherwise generalized pain in the left hip
Lungs:	CTAB	Skin:	No overlying rash
Heart:	Tachycardic, RRR, no M/R/G	Neuro	Decreased sensation and strength in left leg, otherwise no FND

TT	Pt Rm Time	Bed	Patient	Age	ESI	Temp	BP	HR	SpO2	RR
4:16	00:15	Room 22	Marcus (M)	35	3	37.9	142/87	104	96%	20

Clinical Pearls from the Experts: Dr. Vichinski, Dr. Hagar, and Dr. Thompson

- Do not fall for the cell phone sign. Pain is ever-present for a person living with SCD and reacting to it as any person experiencing the same level of pain is perceived as:
  - Aggressive, Confrontational, and Loud
  - A phone is taught as a means of distraction therapy, a means of interim pain management.
- Point tenderness is something scary and needs to be looked at a lot closer.
  - The majority of patients will describe their pain as sharp but also generalized in a specific area; pain at one spot often has a problematic cause, get some imaging.
- Always ask and worry about new focal neurological deficits. Often times these are not the main focus of the pain, however they are important to document and follow up on.
  - Up to 39% of kids under the age of 18 have cerebral infarction, 11% have clinically apparent strokes by the age of 20 and 50% of people by the age of 30 at a minimum have silent cerebral infarctions.



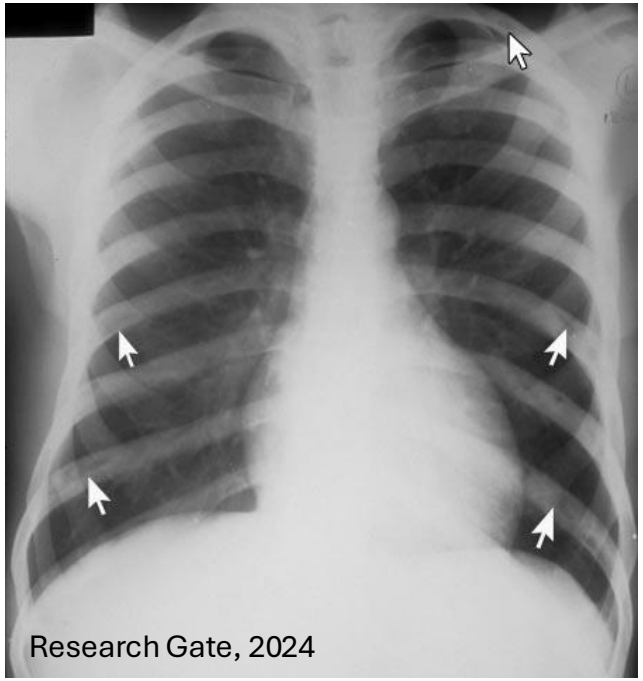
9 y/o M living in St. Louis

Luis A, Blood 2009

Case Presentation: Marcus - 35M

Labs and Imaging:

- CBC: WBC 18 / Hgb 7.2 / Hct 21.6% / Platelets 620
- Retic Count: 12%
- BMP: Na 138 / K 3.9 / Cl 110 / CO2 23 / BUN 9 / Cr 1.1 / Glucose 110
- Urinalysis: mild hematuria, moderate proteinuria, and low specific gravity (hyposthenuria)
- CXR: mild cardiomegaly, no atelectasis or consolidation, multiple small rib infarcts


Research Gate, 2024

TT	Pt Rm Time	Bed	Patient	Age	ESI	Temp	BP	HR	SpO2	RR
5:31	1:30	Room 22	Marcus (M)	35	3	38.2	142/87	87	94%	20

Clinical Pearls from the Experts: Dr. Vichinski, Dr. Hagar, and Dr. Thompson

- CBC: it is very normal for a WBC count to be high, be careful with acute drops in Hgb > 1 from **baseline**, platelets are often higher due to asplenia and overactivation. Lower platelet counts < 200, are very poor prognostic indicators.
  - Don't forget, Anemia is Anemia!
- What is a reticulocyte count and why is it important?
  - The retic count refers to the number of immature red blood cells in a person's blood.
    - High retic count suggests increased RBC destruction (hemolysis) and can indicate a sickle cell crisis, **but it is not diagnostic of a pain crisis**!
    - Low retic count can indicate a problem with the bone marrow's ability to produce new RBCs and indicate an aplastic crisis.
- BMP and UA: Do not always trust the Creatinine, 30% of adults with SCD will develop renal failure. Remember, hyperfiltration (is due to chronic sickling causing reduced blood flow and subsequent compensatory vasodilation raising renal blood flow) hides the failure.
- CXR: we often see relatively normal CXRs, particularly for acute chest syndrome, which is often rapid in onset…quiet before the storm

Case Presentation: Marcus - 35M

- Marcus received 2 rounds of pain management over the course of 4 hours, his physician refused to give him 2 mg of Dilaudid, always giving 1 mg.
  - Patient is sleeping but easily arousable and has been yelling at the nurse, asking for his pain meds. He refuses to wear his oxygen mask and is insisting that his pain is "still a 10." His girlfriend is lying on the bed with shoes on and requests a bus token to go home.
- Going into the third round of pain management, Marcus continued asking for pain medication, describing new chest pain and discomfort. New vitals below.
  - Patient is complaining of new chest pain, asking for more pain meds. Cussing at the nurse.
- Due to this "behavior"…Marcus was asked to leave the emergency department; it was documented that he had oral pain medications at home.

TT	Pt Rm Time	Bed	Patient	Age	ESI	Temp	BP	HR	SpO2	RR
8:31	4:30	Room 22	Marcus (M)	35	3	38.7	142/87	92	92%	23

Case Presentation: Marcus - 35M

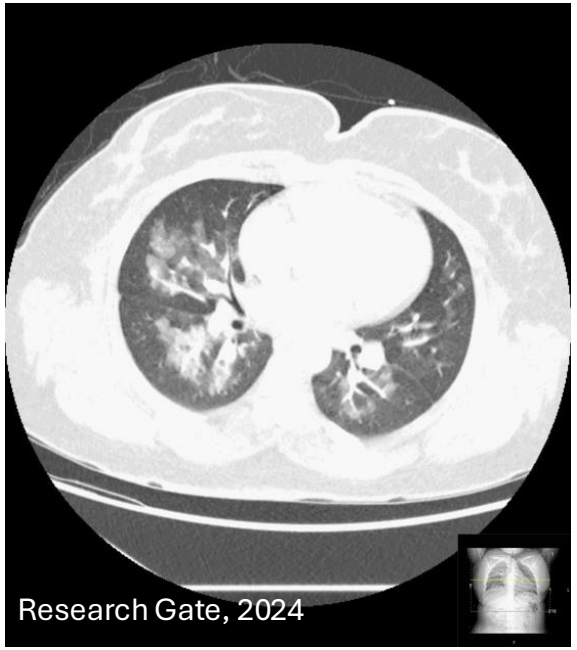
- 2 Days Later: Marcus was brought into the emergency department by EMS. His girlfriend called the ambulance.
  - A: unresponsive, B: crackles appreciated bilaterally, C: hypotensive and tachycardic/weak distal pulses. GCS of Eye 1 Verbal 1 Motor 3; total 5.
  - Patient successfully intubated. POC Hgb 5.8, Glucose 92, and intranasal Narcan administered
  - Patient transfused 2 units of pRBCs and IV fluids
  - Started on Ceftriaxone and Vancomycin
  - Central and ART line successfully placed; patient started on Levophed
  - MICU on its way to evaluate

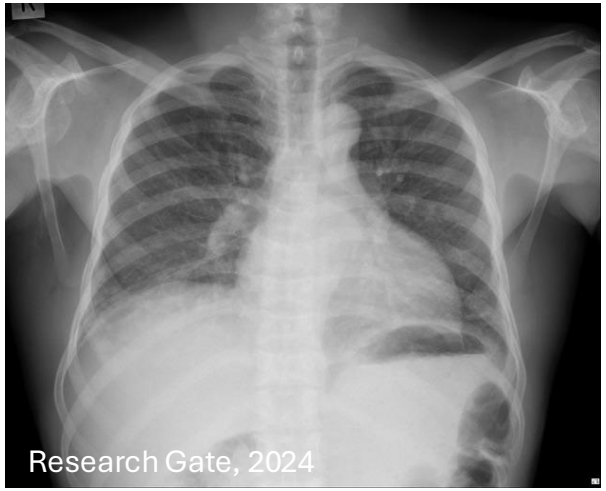
TT	Pt Rm Time	Bed	Patient	Age	ESI	Temp	BP	HR	SpO2	RR
00:01	00:01	Trauma 1	Marcus (M)	35	1	39.2	87/42	142	87% (5L NC)	23

Case Presentation: Marcus - 35M

Labs and Imaging:

- CBC: WBC 24 / Hgb 5.7 / Hct 17.1% / Platelets 190
- Retic Count: 12%
- BMP: Na 138 / K 5.4 / Cl 118 / CO2 10 / BUN 24 / Cr 1.5 / Glucose 92
- CXR: enlarged cardiac silouhette, bilateral lower zone atelectasis w/hazy airspace opacities. Bones diffusely sclerotic.
- CT scan: w/o PE, bilateral perihilar ground glass opacities and consolidation R > L.





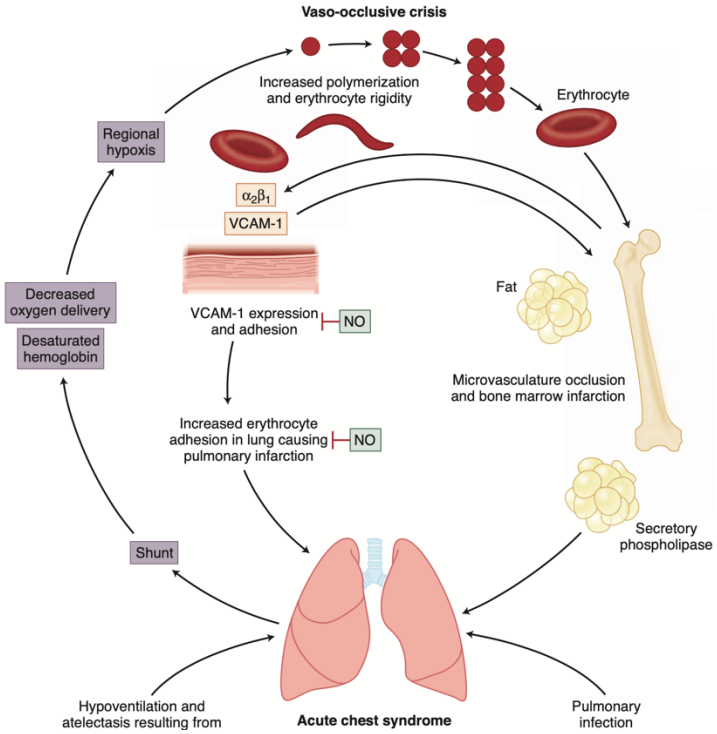
TT	Pt Rm Time	Bed	Patient	Age	ESI	Temp	BP	HR	SpO2	RR
0:34	0:34	Trauma	Marcus (M)	35	3	38.2	142/87	87	94%	20

Vaso-Occlusive Pain Crisis

Vaso-Occlusive Crisis (VOC): A hallmark of SCD characterized by:	• Acute, severe pain due to tissue ischemia • Caused by sickled red blood cells obstructing microvascular blood flow • Can affect any body part, commonly long bones, chest, and abdomen • No reliable objective indicators; patient self-report is the gold standard
Key Points for Emergency Management:	• Prompt, aggressive pain management • Appropriate hydration • Identify potential triggers (e.g., infection, dehydration) • Assess for complications (e.g., acute chest syndrome)
Laboratory Findings:	• Leukocytosis > 20,000/µL with left shift may indicate infection • Mild elevations in bilirubin and LDH are common • Hemoglobin levels may be lower than baseline • Increased reticulocyte count
Critical Reminders:	• Patient's self-reported pain is the primary diagnostic tool • Behavioral cues or lab values do not reliably indicate pain severity • Early, adequate analgesia is crucial for optimal outcomes

Amanda M, ASH 2020
Duroseau Y, JACEP 2021
Ojo AS, J Clin Med Res. 2023

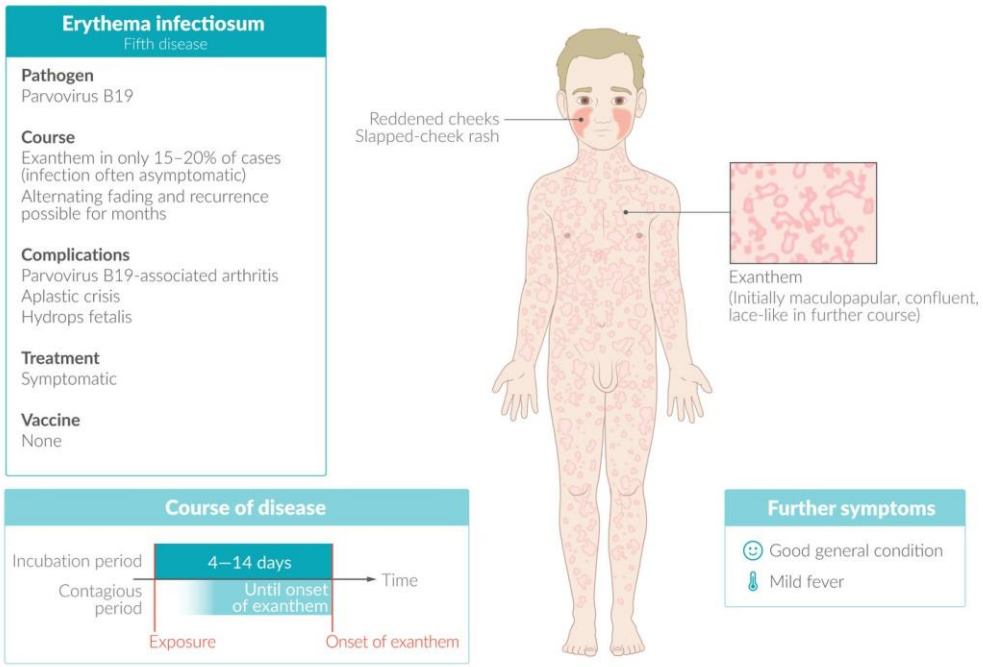
Acute Chest Syndrome



Acute Chest Syndrome (ACS) is a severe complication of sickle cell disease characterized by:	• New pulmonary infiltrate on chest X-ray with at least one additional symptom: fever >38.5°C, cough, wheezing, tachypnea, or chest pain • Leading cause of death in patients with HbSS • Often develops 1-3 days after hospitalization for acute pain crisis
Key Points for Emergency Management:	• Prompt, aggressive pain management and appropriate hydration • Early administration of broad-spectrum antibiotics • Supplemental oxygen to maintain saturation ≥95% • Consider blood transfusion in consultation with hematology
Ancillary Tests:	• Ancillary Tests: CBC w/diff, retic count, LFTs, LDH, BMP, UA, CXR, and Blood Cx's • Up to 30% of cases are due to infection most common is Chlamydia pneumoniae (in **adults**) and Mycoplasma pneumoniae (in **pediatrics**).
Critical Reminders:	• Patient's self-reported pain is the primary diagnostic tool • No reliable objective indicators exist for acute chest syndrome • Early recognition and treatment are crucial for optimal outcomes • Monitor closely for rapid clinical deterioration

Alghamdi FA, Sci Rep 2024
Vichinsky EP, NEJM 2000
Castro O, Blood 1994

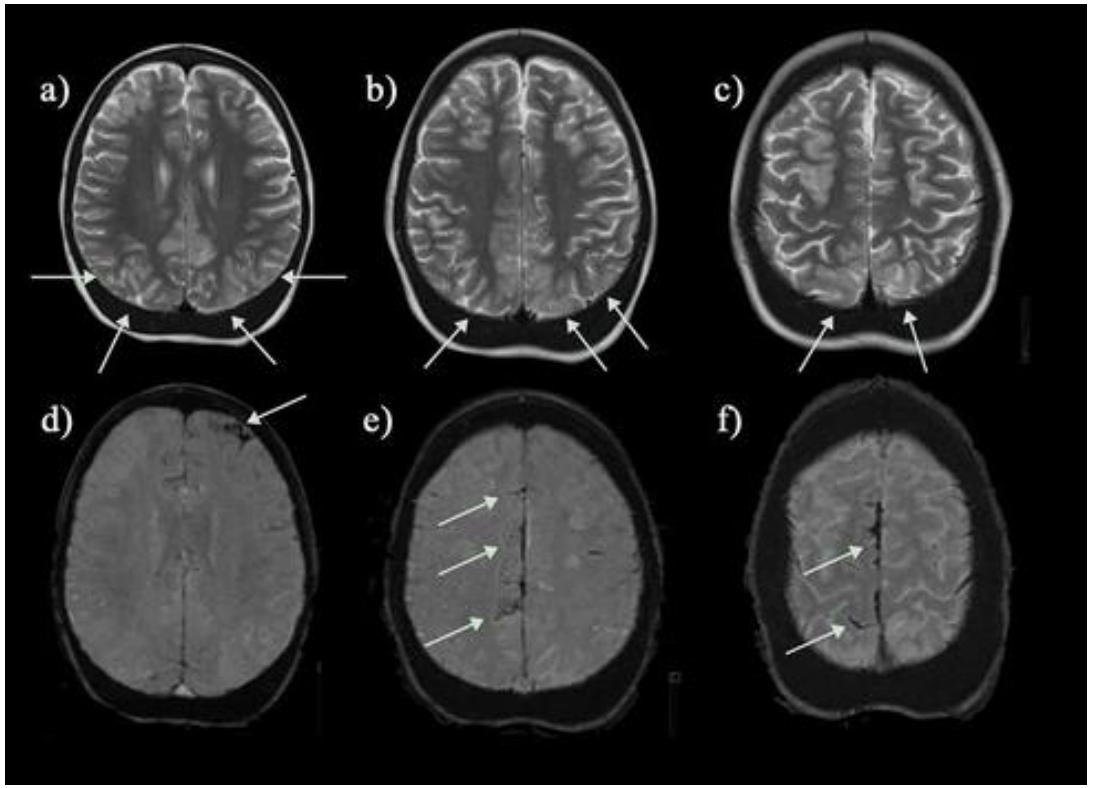
Aplastic Crisis: Why the retic count is so important!



Aplastic Crisis in Sickle Cell Disease	• Sudden cessation of red blood cell production in the bone marrow • Rapid decrease in hemoglobin with reticulocytopenia • Most commonly caused by human parvovirus B19 infection • Typically affects children with sickle cell disease
Key Points for Emergency Management:	• Recognize the signs: sudden severe anemia, fatigue, shortness of breath, and potentially hemodynamic compromise • Obtain immediate laboratory tests: CBC with reticulocyte count, hemoglobin level, and parvovirus B19 testing • Initiate prompt red blood cell transfusion to address severe anemia • Provide supportive care, including close monitoring and potential hospitalization for severe cases
Clinical Presentation:	• Symptoms of severe anemia (fatigue, weakness, shortness of breath) • Pale skin or lips, possible erythema infectiosum • Fever and flu-like symptoms • Rapid heart rate
Key Lab Findings:	• Key Laboratory Findings:Hemoglobin drops below baseline (usually 6-9 g/dL) • Reticulocyte count falls below 100,000/µL (normal range 5-15%) • Positive test for parvovirus B19

Serjeant GR, Lancet 1981
Rees DC, J of Haemotology 2003

Acute Stroke



Epidemiology and Risk	• Stroke is a leading cause of death in SCD, accounting for up to 10% of deaths • Lifetime risk of overt stroke is 25-30% in SCD patients • Children with SCD have 333 times greater stroke risk than healthy children • Up to 39% of children develop silent cerebral infarcts by age 18
Types and Timing	• Both ischemic and hemorrhagic strokes occur • Ischemic strokes more common in children • Hemorrhagic strokes more common in adults with SCD
Key Points for Emergency Management:	• Prompt blood transfusion within 2 hours of symptom onset • tPA can be safely administered if patients qualify within 4.5 hours • Type of transfusion (simple, modified exchange, or apheresis) depends on individual factors • Requires coordinated care between hematology and neurology
Critical Reminders:	• Critical Reminders:Early recognition is crucial for optimal outcomes • Maintain high clinical suspicion in SCD patients with neurological symptoms • Treatment decisions should not delay blood transfusion therapy • Regular screening with transcranial Doppler recommended for prevention (especially in younger patients)

De Baun MR, ASH 2020
Adams RJ, Stroke 2017

Clinical Pearls from the Experts: Dr. Vichinski, Dr. Hagar, and Dr. Thompson

- Blood is the way to go!
  - While published data remains limited, there is a growing body of expert consensus and ongoing studies supporting the critical role of early exchange transfusion or transfusion in recovery, particularly for the brain (validated), heart, and lungs.
  - Leading Sickle Cell centers strongly recommend transfusion when acute chest syndrome is suspected, underscoring its pivotal role in improving patient outcomes.

Insights gathered from expert discussions underscore a critical truth:
“Sickle cell disease in the United States is more than a genetic condition; it is a construct shaped by systemic racism, social inequities, and structural barriers that magnify its impact on Black lives. While the disease itself is a manageable condition—one that many can live with and survive—what proves most lethal are the inequities in access to care, resources, and support. It is not the sickling alone that kills but the compounded effects of neglect, discrimination, and the failure to prioritize equitable healthcare. Sickle cell disease, therefore, reveals the devastating interplay between biology and the burden of societal injustice.”

Case Presentation: Marcus - 35M

- 2 Days Later: Marcus was brought into the emergency department by EMS. His girlfriend called the ambulance.
  - A: unresponsive, B: crackles appreciated bilaterally, C: hypotensive and tachycardic/weak distal pulses. GCS of Eye 1 Verbal 1 Motor 3; total 5.
  - Patient successfully intubated. POC Hgb 5.8, Glucose 92, and intranasal Narcan administered
  - Patient transfused 2 units of pRBCs and IV fluids
  - Started on Ceftriaxone and Vancomycin
  - Central and ART line successfully placed; patient started on Levophed
  - MICU on its way to evaluate

TT	Pt Rm Time	Bed	Patient	Age	ESI	Temp	BP	HR	SpO2	RR
00:01	00:01	Trauma 1	Marcus (M)	35	1	39.2	87/42	142	87% (5L NC)	23

Case Presentation: Marcus - 35M

Timeline:

- Marcus was transferred in “stable” condition to the MICU
- Marcus passed away 2 days later due to multi-organ failure caused by acute chest syndrome
- On autopsy, he was found to have a bone marrow infarction w/possible osteomyelitis in the right femur.
  - His lungs were also found to have lipid-laden macrophages in the bronchoalveolar lavage fluid, indicative of fat embolism.
- Although infection is the most common cause, as previously stated, 9% - 11% of acute chest syndrome is caused by bone marrow/fat emboli.

Take home points:

- **The Gold Standard for identifying a Sickle Cell Pain Crisis is a patient's voice**
- Words really do matter and "sickler" is not one that should be used.
- Important questions:
  - Is this your typical pain?
  - Do you have a pain plan? What are some medications that work best for you?
- We can all be hydroxyur-advocates!
- New focal neurological deficits are common and very important, even in patients as young as 10!
- For brain, heart, and chest, consult your hematologist and consider blood!
- Point tenderness is not normal, I would get some imaging!

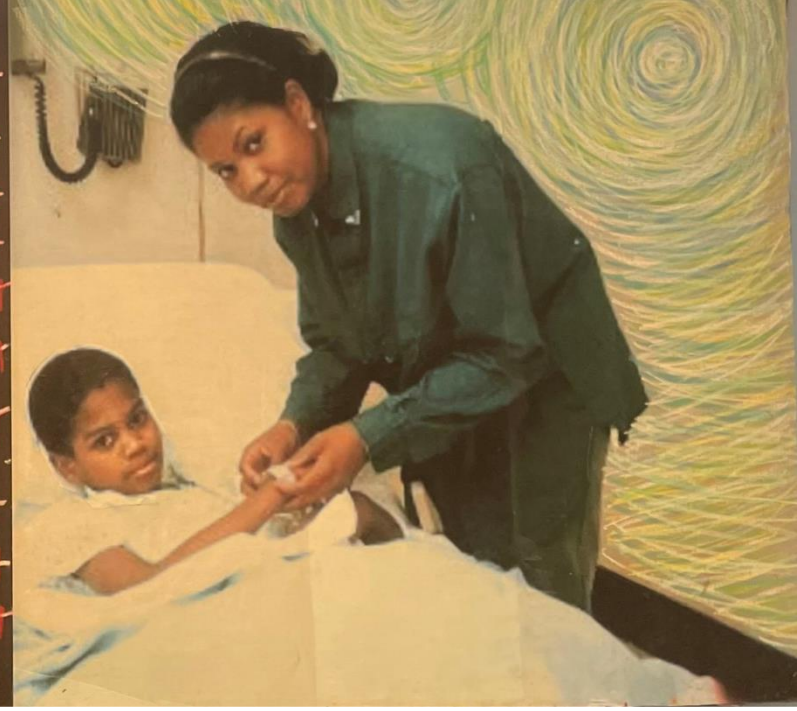
Thank You Both

It's the pain of a broken spirit that hurts more than the physical pain. It's the pain of not being believed at school, at work. It's the pain of not being believed at the hospital. It's the pain of the choice to stay at home and suffer, rather than go to a local hospital and be treated like an addict. That's what breaks you.

- Hertz Nazaire

October 2nd, 1973 - October 29th, 2021

Passed away from complications due to Sickle Cell Disease at the age of 48



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A Brief Conversation about Sickle Cell Disease with Adrienne Shapiro

Adrienne Shapiro is the co-founder and science administrator for Axis Advocacy, an organization dedicated to advancing efforts to cure sickle cell disease. As the fourth generation of mothers in her family to have a child born with this condition, Adrienne's life has been shaped by her unwavering commitment to improving healthcare and quality of life for those affected. A fierce advocate, she believes deeply in the power of science and has emerged as a prominent voice in the fight for equitable care and groundbreaking treatments.

Adrienne was an early supporter of transformative therapies, including bone marrow transplants and stem cell research pioneered by Dr. Kohn. Over the past three years, she has amplified her role as a stem cell activist, championing funding for medical trials through CIRM (California Institute for Regenerative Medicine). Adrienne speaks nationally and internationally at educational conferences, attends key seminars, and actively lobbies Southern California Congress members for initiatives like the Sickle Cell Education Act.