CMSS Council of Medical Specialty Societies
CMSS Annual Meeting 2022
Specialty Societies: Stronger Together
November 9-11, 2022
Washington, DC

Emergency Department Sickle Cell Care Coalition: Working Together to Improve Care

EDSC3
Emergency Department Sickle Cell Care Coalition

Caroline Freiermuth, MD, MHS
Chair, EDSC3
Associate Professor of Emergency Medicine
University of Cincinnati College of Medicine

David Brousseau MD, MS
Co-Chair, EDSC3
Professor of Pediatrics
Medical College of Wisconsin

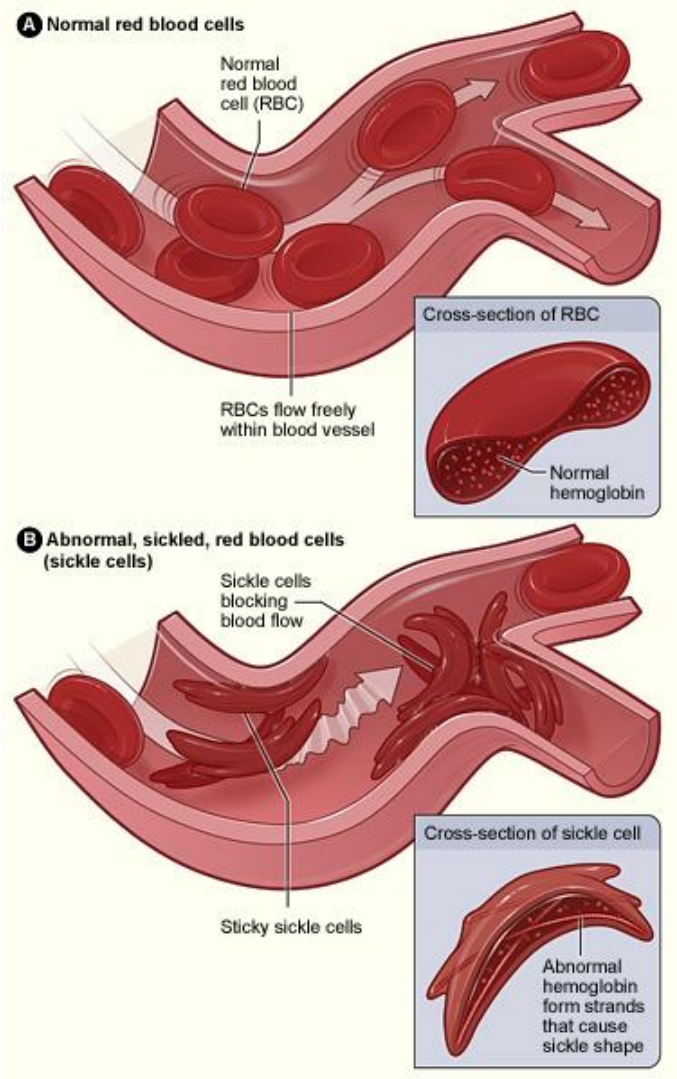
Patricia Kavanagh, MD
Immediate Past Chair, EDSC3
Associate Professor of Pediatrics
Boston University School of Medicine

Disclosures

- American College of Emergency Physicians
  - Registration and travel expenses related to this conference paid on behalf of Dr. Freiermuth
  - Information technology support for sickle cell point of care tool
  - Administrative support for EDSC3
- Novartis
  - Costs associated with development of point of care tool
    - No input regarding content
- Pfizer
  - Unrestricted grants for quality improvement projects

Background

- 90-100,000 people living with sickle cell disease (SCD) in the USA
  - Mainly those of African descent
- Abnormal cells cause vaso-occlusion
  - Pain is most common complication
    - Unpredictable
    - Intense



Wikipedia.com

Medical Model

Barriers to Optimal Care

- Emergency provider lack of education
- Bias
  - Racial
  - Disease
  - Patients labelled as drug seekers or frequent fliers
- Crowding

Impact on Care

- Longer wait times for pain medication
- Variable care
- Mistrust between providers and patients
- Avoidance of the emergency department
- High rates of readmission

Emergency Department Sickle Cell Care Coalition

- 2015 initial meeting
- Impetus driven by patients
- Organized by Dr. Patricia Kavanagh
- Mission: to promote ***evidence-based emergency care*** and ***optimize provider-patient-family communication*** in the delivery of the emergency care for patients with SCD

Who We Are

EDSC³ Emergency Department Sickle Cell Care Coalition

Professional Organizations	Federal Partners	Community Groups
American College of Emergency Physicians	Centers for Disease Control and Prevention	Sickle Cell Disease Association of America
Emergency Nurses Association	Health Resources and Services Administration	Sickle Cell Community Consortium
American Society of Hematology	National Heart, Lung and Blood Institute	Minimum of 2 individuals living with SCD
American Society of Pediatric Hematology/Oncology	The Joint Commission	
American Academy of Pediatrics		
American Society of Health-System Pharmacists		

Making Sickle Cell Disease an ED Priority

Areas of Focus

Accomplishments-Education

- Lectures to emergency clinicians and nurses through national, regional, and institutional meetings
- Emergency Nurses Association Topic Brief Sept 2021
- Webinars in conjunction with CDC and NHLBI
- Twitter chats, hosted by the American Society of Health-System Pharmacists

Accomplishments-Research

- Created funding opportunities for quality improvement research
  - Sponsored by Pfizer, administered by the American College of Emergency Physicians
  - 6 sites
- Emergency Medicine Practice Research Network survey looking at attitudes towards patients
- Annals of Emergency Medicine Supplement on Sickle Cell Disease
  - September 2020
  - Funded by the Department of Health and Human Services, Office of Minority Health

Accomplishments- Advocacy

- Supported ASH in requesting additional funding to expand the SCD Data Collection Program
- Emergency Nurses Association Resolution 2019- 'Management of Vaso-Occlusive Episodes in Persons with Sickle Cell Disease in the Emergency Department'
- Bolstered multi-center application to HRSA for Sickle Cell Disease Treatment and Demonstration program

Accomplishments- Community Outreach

- Steering committee members of Sickle Cell Disease Coalition
- Present at multiple patient care organization conferences each year
- Maintain website with links to SCD resources

Point of Care Tool Development

- Stakeholders all reported need for guidance
  - Summit August 2018
- Emergency physicians want a voice in guidelines/recommendations regarding the care they deliver
  - Lean on emergency medicine society communication and website for information
- Iterative process

Communication

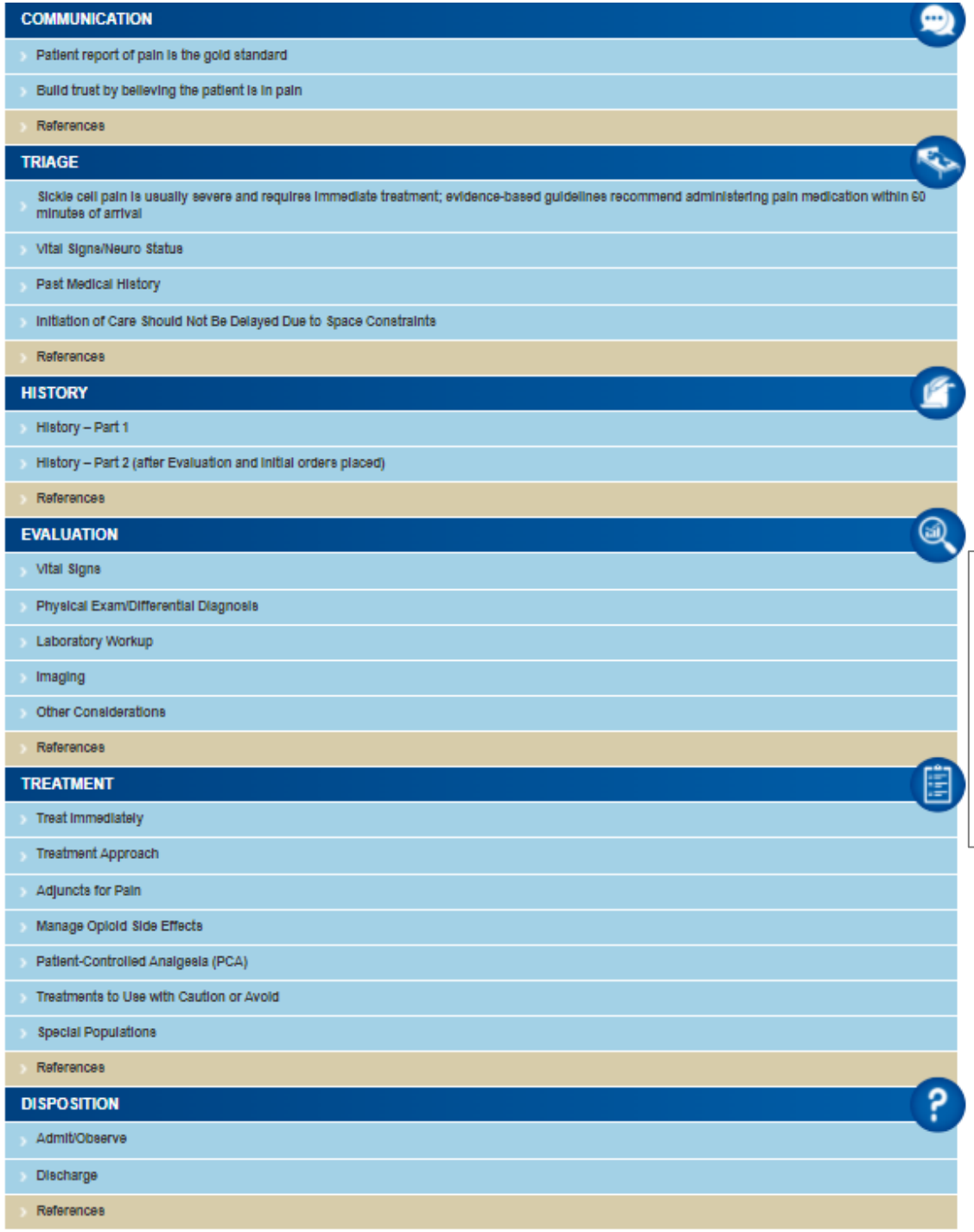
Triage

History

Evaluation

Treatment

Disposition



FREE!

https://www.acep.org/sickle-cell/

American College of Emergency Physicians
ADVANCING EMERGENCY CARE

American Society of Hematology

COMMUNICATION

- Patient report of pain is the gold standard
- Build trust by believing the patient is in pain
- References

TRIAGE

- Sickle cell pain is usually severe and requires immediate treatment; evidence-based guidelines recommend administering pain medication within 60 minutes of arrival
- Vital Signs/Neuro Status
- Past Medical History
- Initiation of Care Should Not Be Delayed Due to Space Constraints
- References

HISTORY

- History – Part 1
- History – Part 2 (after Evaluation and initial orders placed)
- References

COMMUNICATION

Patient report of pain is the gold standard

There are no vital sign changes or lab values that confirm or rule-out a sickle cell pain crisis

Do not refer to patients with SCD as "sicklers," as this is a derogatory term

Requests for specific pain medicines/doses are most commonly due to past experience, not drug-seeking behavior

- Opioid use was stable from 2008-2013 in the SCD population, in contrast to the general US population
- Deaths from opioid overdose was ≤ 10 per year in individuals with SCD from 1999-2013, representing only 0.77% of all deaths in this population, significantly lower than other non-cancer conditions including low back pain, fibromyalgia and migraine

Build trust by believing the patient is in pain

Patients may not be glad to see you – show them you are here to help

- Negative ED experiences in the past may make them guarded or mistrustful
- Pain can make anyone irritable, impatient, or upset
- Empathetic nonverbal communication is essential (eye contact, facial expression, gestures)
- Be patient when asking questions; it is often difficult to speak when in severe pain

Patients may bring a caregiver with them, as it may be difficult to understand treatment plans and ask questions when in severe pain.

EVALUATION

- Vital Signs
- Physical Exam/Differential Diagnosis
- Laboratory Workup
- Imaging
- Other Considerations
- References

TREATMENT

- Treat Immediately
- Treatment Approach
- Adjuncts for Pain
- Manage Opioid Side Effects
- Patient-Controlled Analgesia (PCA)
- Treatments to Use with Caution or Avoid
- Special Populations
- References

DISPOSITION

- Admit/Observe
- Discharge
- References

Physical Exam/Differential Diagnosis

NOTE: Severe SCD complications often develop during a pain crisis or mimic one. Contact hematology EMERGENTLY if you suspect one, such as acute stroke or acute chest syndrome with significant respiratory distress.

Location of pain

- Localized bony pain that is different from previous pain crises
  - Osteomyelitis/septic arthritis
  - Priapism (sometimes report thigh pain)
  - Avascular necrosis
- Calf pain could be DVT
- Diffuse pain could be systemic infection: Check vital signs, look for rash or petechiae on skin (including palms/soles) and gums

Altered mental status, severe or atypical headache, focal neurologic findings (such as vision changes), or new seizure: Consider stroke in children and adults

Fatigue/jaundice/pallor: Consider aplastic anemia, sepsis or hemolysis due to delayed hemolytic transfusion reaction

Chest pain: Consider acute chest syndrome (leading cause of death in SCD)

Respiratory distress: Consider acute chest syndrome, pulmonary embolism and sepsis

Abdominal pain or distension: Assess for splenomegaly or hepatomegaly (splenic or hepatic sequestration), RUQ pain (acute cholecystitis), and consider priapism if low abdominal pain

Assess fluid status for dehydration

Suggested Parenteral Opioid and NSAID Doses for Adults and Adolescents (≥ 12 years or ≥ 50 kg)

Pain Medication	Dose	Max Single Dose	Frequency
Morphine IV*	0.1 mg/kg	10 mg	Repeat every 15-30 minutes until pain controlleds
Hydromorphone IV	0.015 mg/kg	4 mg	Repeat every 15-30 minutes until pain controlled
Ketorolac IV*	0.5 mg/kg	15 mg	Once

***Use with caution in patients with mild renal dysfunction**

Adjuncts for Pain

NSAIDs: Can be given in addition to opioids to manage acute pain and treat the inflammation associated with a sickle cell crisis

- Exclusions include patients with significant renal dysfunction (up to 30% of adults with SCD) and full-dose anticoagulation; use caution in patients with peptic ulcer disease

IV fluids: No evidence that IV fluids are beneficial in euvolemic patients; may cause fluid overload and acute chest syndrome in certain patients

- Consider IVF infusion rather than bolus and administer less than maintenance fluids if able to drink (e.g., 75% maintenance)

Non-pharmacologic interventions: Heat packs (no ice), blankets, distraction (using phone, watching TV)

Manage Opioid Side Effects

Pruritis:

- Oral diphenhydramine 25 mg recommended. Avoid IV diphenhydramine push because it worsens sedation.
- Low dose naloxone infusion (1 mcg/kg/hr) does not reverse effects of opioids; can be titrated up 0.5 mcg/kg/hr every 3-4 hours, maximum 4-5 mcg/kg/hr
- IV diphenhydramine 25 mg if alternatives listed above do not control pruritis or are unavailable

Nausea/vomiting: Ondansetron is commonly used; use caution with prochlorperazine and metoclopramide due to sedating effects

Respiratory depression: Naloxone 0.4 mg/dose IV to alleviate sedation (not reverse analgesia)

Patient-Controlled Analgesia (PCA)

Treatments to Use with Caution or Avoid

Steroids: Can trigger severe pain crises, but short courses can safely treat select conditions (e.g., asthma) when done in coordination with hematology/SCD provider

Ketamine: Guidelines found insufficient evidence for use outside of the inpatient setting

Blood transfusions: Do not give transfusions to manage sickle cell pain crisis unless directed by the patient's hematologist or SCD provider

Oxygen: Do not use oxygen unless patient is hypoxic compared to their usual oxygen level

DISPOSITION

Admit/Observe

Discharge

If patient believes they can manage pain at home

Recommend close follow-up with hematology/SCD provider. Refer to a specialist if not already established

- Recognize that there is not always ready access to a hematologist. Efforts should be made to establish a referral pathway to a local physician to manage these patients

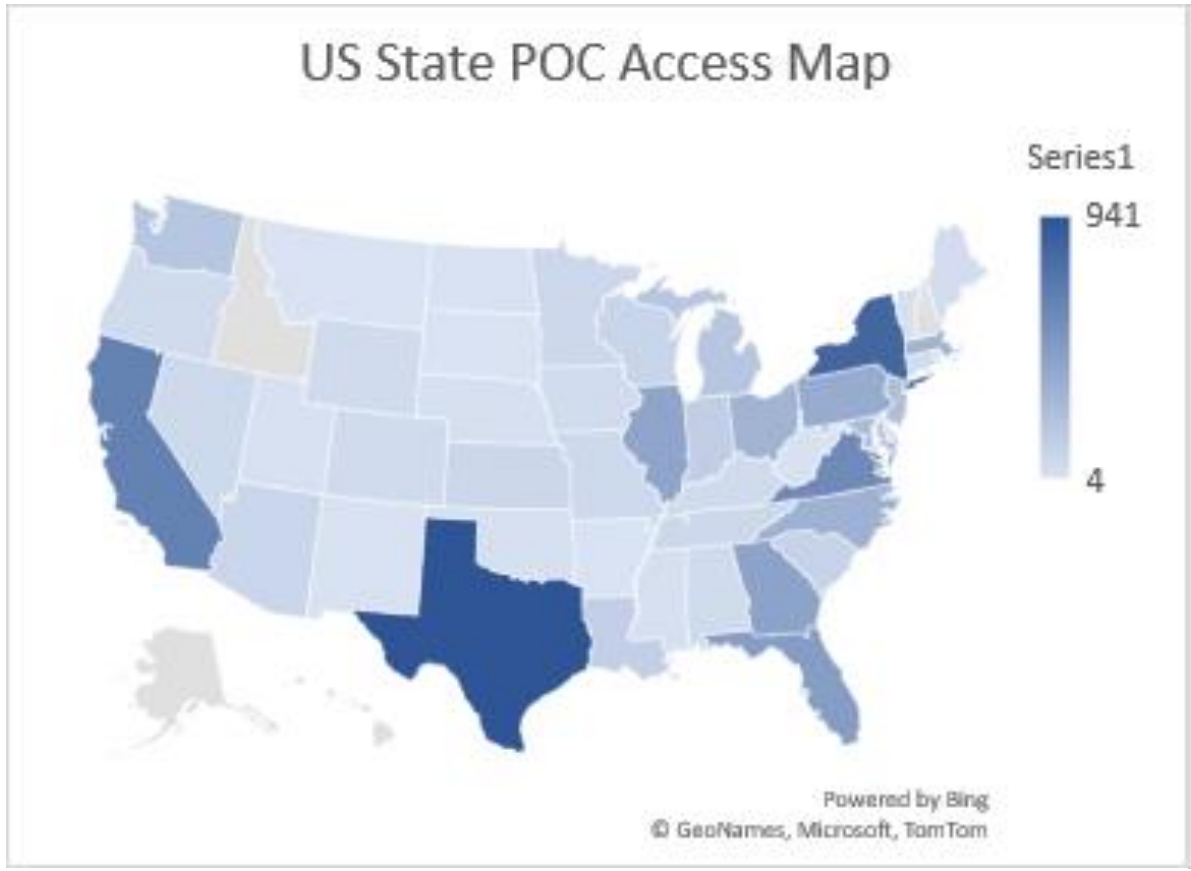
Ensure adequate access to medications for management of pain as an outpatient

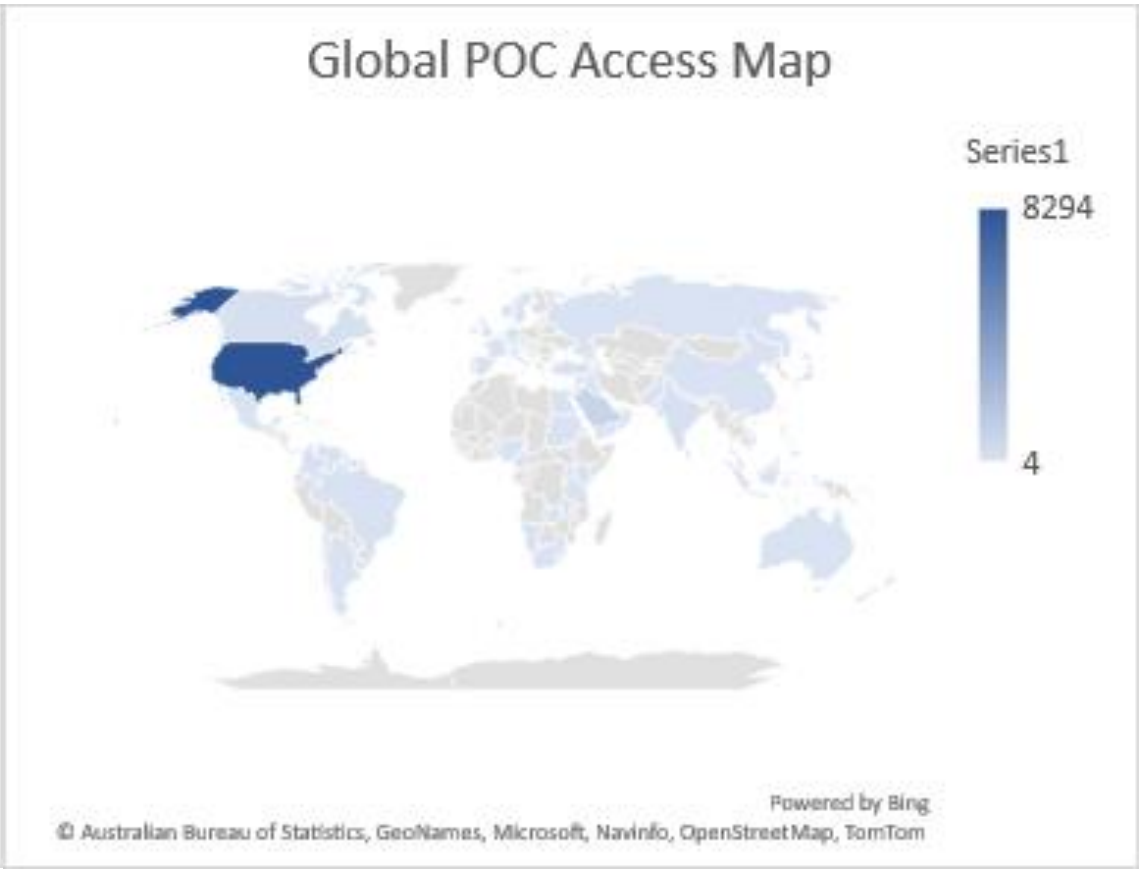
- o Opioids:
  - Short-term (e.g., 3 days) opioid prescription as bridge to next outpatient visit
  - If concern for opioid misuse/overdose – contact outpatient provider to make plan
- Non-opioids including acetaminophen, topical or oral NSAID (if not contraindicated): Continue for short-course (5-7 days total)
- Non-pharmacologic means of pain control: Continue heat, hydration, distraction; avoid using ice
- Bowel regimen to avoid opioid-induced constipation

Give clear return precautions, such as fever, difficulty breathing, chest pain, changes in mental status, uncontrolled pain

Some patients with frequent visits have unmet social or behavioral health needs including lack of insurance or transportation, unable to pay for prescriptions, unstable housing, etc. Make referrals to social work or case management as appropriate. Learn more here.

Point of Care Tool Metrics





Where do we go from here?

- Dissemination
  - Education
  - Point of care tool
  - Grow EM network to contribute to the mission of EDSC3

- Must measure impact
  - Define outcome measures
  - Utilize existing research networks

Questions?

- Caroline Freiermuth: caroline.freiermuth@uc.edu
- David Brousseau: dbrousse@mcw.edu
- Patricia Kavanagh: patricia.Kavanagh@bmc.org
- EDSC3: edsc3@acep.org

- EDSC3: https://www.acep.org/by-medical-focus/hematology/sickle-cell/
- Point of Care Tool: https://www.acep.org/patient-care/sickle-cell/

References

1. Glassberg JA. Improving Emergency Department-Based Care of Sickle Cell Pain. Hematology Am Soc Hematol Educ Program. 2017 Dec 8;2017(1):412-417.
2. Haywood C Jr, Tanabe P, Naik R, Beach MC, Lanzkron S. The impact of race and disease on sickle cell patient wait times in the emergency department. *Am J Emerg Med*. 2013;31(4):651-656.
3. Abdallah K, Buscetta A, Cooper K, Byeon J, Crouch A, Pink S, Minniti C, Bonham VL. Emergency Department Utilization for Patients Living With Sickle Cell Disease: Psychosocial Predictors of Health Care Behaviors. Ann Emerg Med. 2020 Sep;76(3S):S56-S63.