

Developing an Emergency Department Order Set for Sickle Cell Disease Acute Pain

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Background



- Sickle cell disease (SCD) is a life-threatening, multifaceted, debilitating disease
- Recurrent vaso-occlusive episodes (VOEs) are the hallmark of SCD
 - Patients with SCD have 2.59 emergency department (ED) visits per year on average, the majority of which are for VOEs^{2,3}
 - Inequity in ED care exists⁴⁻⁶
- Guidelines recommend treatment within 30 minutes of triage, but this is rarely achieved in practice
- There has been some success with implementing ED protocols to manage VOEs in both adults and children with SCD^{3,7,8}

Objective



- **Our goal was to develop an evidence-based order set** that could be implemented in New York City (NYC) EDs to expedite and standardize emergency care for SCD patients presenting with acute pain
 - Similar to the Community Care of North Carolina Sickle Cell Task Force local protocol⁹
- Improve the quality and consistency of care provided to patients with SCD

Used a RAND/UCLA modified Delphi panel method



- A valid, reliable, and reproducible method that can be used to generate consensus
- Convened 10 clinicians practicing in NYC with an average of 11 years' experience caring for patients with SCD
- Provided clinicians with a review of evidence primarily based on the National Heart, Lung, and Blood Institute (NHLBI) guidelines on how to best manage SCD pain in the ED



5 emergency medicine



2 emergency medicine
& internal medicine



2 hematology



1 pain & palliative care

Rated 202 items that could be included in an order set



A. Triage (e.g., initiate SCD protocol, assign ESI level 2)

B. Initial medical encounter (e.g., implement individualized plan)

C. Targeted evaluation (i.e., rule out other complications)

D. Initial pain management

E. First pain reassessment

F. Second pain reassessment

G. Third pain reassessment

H. Preventive care (e.g., vaccinations, referrals)

I. Discharge (e.g., prescriptions, follow-up appointments)

J. Other considerations (e.g., non-pharmacologic approaches)

Consensus order set



A. TRIAGE	
<ul style="list-style-type: none"><input type="checkbox"/> Identify SCD patient and initiate SCD protocol<input type="checkbox"/> Assess vitals, including oxygen saturation (O₂ sat)<input type="checkbox"/> Assess pain using VAS or verbal scale (1-10)ⁱ<input type="checkbox"/> Confirm allergies to medications (opiates, NSAIDS, antibiotics, etc.)<input type="checkbox"/> Assign ESI level 2<input type="checkbox"/> Begin implementation of rapid protocol (initiate analgesic therapy <30 minutes after triage)	
B. INITIAL MEDICAL ENCOUNTER	
<ul style="list-style-type: none"><input type="checkbox"/> Review vitals (including O₂ sat)<input type="checkbox"/> Assess pain using VAS or verbal scale (1-10)<input type="checkbox"/> Note treatment prior to coming to ED or in triage (opioids, NSAIDS)<input type="checkbox"/> Note baseline hemoglobin*<input type="checkbox"/> Note date of and reaction to last transfusion*	
Assess if patient has a documented SCD treatment plan:	
<ul style="list-style-type: none"><input type="checkbox"/> If yes, review with patient and integrate with items in sections E [INITIAL PAIN MANAGEMENT] and F, G, H [PAIN REASSESSMENTS]<input type="checkbox"/> If no, attempt to find analgesic history during previous ED visits in medical record	
<ul style="list-style-type: none"><input type="checkbox"/> Confirm usual analgesic type and dose with patient	
C. DRAW LABS	
Draw labs as appropriate:	
<ul style="list-style-type: none"><input type="checkbox"/> CBC with differential<input type="checkbox"/> Reticulocyte count<input type="checkbox"/> Electrolytes (CHEM-7)<input type="checkbox"/> ALT and AST*<input type="checkbox"/> LDH	<ul style="list-style-type: none"><input type="checkbox"/> Bilirubin<input type="checkbox"/> Type and screen (if no active type and screen)<input type="checkbox"/> Hemoglobin fractionation/electrophoresis*<input type="checkbox"/> Iron studies (Fe, TIBC, Ferritin) if not performed in the past 90 days

*The majority of the panel rated as likely to improve outcomes, with at least two panelists disagreeing.

Consensus order set



D. PERFORM TARGETED EVALUATION	
Evaluate if patient experiencing their typical VOE symptoms:	
<input type="checkbox"/> If yes, confirm usual analgesic type and dose with patient <input type="checkbox"/> If O ₂ sat <95%, provide oxygen (not indicated if O ₂ sat ≥95%)	
If concerned for PE : <input type="checkbox"/> Confirm adequate renal function <input type="checkbox"/> Order CT angiogram	If concerned for MI : <input type="checkbox"/> Order EKG <input type="checkbox"/> Send troponin
If concerned for acute chest syndrome : <input type="checkbox"/> Order chest radiograph <input type="checkbox"/> Hematology consult, consider: <input type="checkbox"/> Adding hemoglobin fractionation/ electrophoresis to labs <input type="checkbox"/> Exchange transfusion	If concerned for stroke (e.g., patient reports headache): <input type="checkbox"/> Implement SCD stroke protocol, if available <input type="checkbox"/> Order brain imaging <input type="checkbox"/> Hematology consult, consider: <input type="checkbox"/> Adding hemoglobin fractionation/ electrophoresis to labs <input type="checkbox"/> Exchange transfusion
If concerned for worsening anemia : <input type="checkbox"/> Notify blood bank for phenotype matched red cells Add the following to labs: <input type="checkbox"/> CBC with differential <input type="checkbox"/> Type and screen <input type="checkbox"/> Hemoglobin fractionation/electrophoresis <input type="checkbox"/> Reticulocyte count <input type="checkbox"/> LDH <input type="checkbox"/> Total and direct bilirubin <input type="checkbox"/> Iron studies (Fe, TIBC, Ferritin)	If concerned for sequestration or acute cholecystitis (e.g., patient reports abdominal pain): <input type="checkbox"/> Order abdominal ultrasound Add the following to labs: <input type="checkbox"/> CBC with differential <input type="checkbox"/> Type and screen <input type="checkbox"/> ALT and AST <input type="checkbox"/> Total and direct bilirubin
Assess SIRS criteria : T >38°C (100.4°F) or <36°C (96.8°F); HR >90; RR >20 or PaCO ₂ <32mmHg; WBC >12,000/mm ³ , <4,000/mm ³ , or >10% bands	If ≥2 SIRS criteria present: <input type="checkbox"/> Implement sepsis protocol <input type="checkbox"/> Consider empiric treatment

Consensus order set



E. INITIAL PAIN MANAGEMENT: Initiate analgesic therapy within 30 minutes of triage		
<input type="checkbox"/> If the patient has a documented individualized SCD pain plan, integrate here <input type="checkbox"/> If opioid is administered, initiate continuous O ₂ sat monitoring		
If the patient has an opioid allergy , provide alternative (assess renal/liver function as needed):		
For children: <ul style="list-style-type: none"> <input type="checkbox"/> PO acetaminophen 15mg/kg <input type="checkbox"/> PO ibuprofen 10mg/kg <input type="checkbox"/> If >2 years IV ketorolac 0.5mg/kg <input type="checkbox"/> If <2 years IV ketorolac 0.25mg/kg 	For adults: <ul style="list-style-type: none"> <input type="checkbox"/> PO acetaminophen 975mg <input type="checkbox"/> PO ibuprofen 600mg <input type="checkbox"/> IV ketorolac 30mg 	Alternative for adults: <ul style="list-style-type: none"> <input type="checkbox"/> PO acetaminophen 650mg* <input type="checkbox"/> IV ketamine 0.25mg/kg* <input type="checkbox"/> IV ketorolac 15mg
If patient has IV access (e.g., peripheral or central line), administer opioid IV (1 st dose)*:		
For patients who are not opioid naïve: <ul style="list-style-type: none"> <input type="checkbox"/> Calculate and administer patient-specific opioid dose (IV route preferred)ⁱⁱ 	For patients who are opioid naïve or with no available analgesic history, administer: <ul style="list-style-type: none"> <input type="checkbox"/> Morphine 0.1mg/kg^{iii*} <input type="checkbox"/> Hydromorphone 0.02mg/kg^{4*} <input type="checkbox"/> Ketamine 0.25mg/kg^{iv*} Avoid meperidine*	
If patient does not have IV access , administer opioid via other routes (1 st dose):		
For patients who are not opioid naïve: <ul style="list-style-type: none"> <input type="checkbox"/> Calculate and administer patient-specific opioid dose (SQ if no IV access)² 	For patients who are opioid naïve or with no available analgesic history, administer: <ul style="list-style-type: none"> For children: <ul style="list-style-type: none"> <input type="checkbox"/> PO hydromorphone 0.05mg/kg <input type="checkbox"/> PO morphine 0.3mg/kg Avoid IN fentanyl in patients under <7 years old or <10kg⁵ 	For adults: <ul style="list-style-type: none"> <input type="checkbox"/> SQ morphine 0.1mg/kg <input type="checkbox"/> SQ hydromorphone 0.02mg/kg <input type="checkbox"/> PO morphine 30mg* <input type="checkbox"/> PO hydromorphone 5mg <input type="checkbox"/> IN fentanyl 2-3 doses 5 minutes apart (max single dose (100µg) may limit efficacy, especially >65kg)^v

Consensus order set



F. FIRST PAIN REASSESSMENT: Within 30 minutes (60 minutes after triage)

Assess pain using VAS or verbal scale (1-10):

If VAS ≥ 5 :

If no hypoxia or sedation:

- Repeat initial dose of IV opioid (2nd dose) if route is available (if route is not available, consider other routes)
- Escalate initial dose of IV opioid by 25%

If signs of excessive sedation:

- Decrease dose of IV opioid

If VAS ≤ 4 see **G [SECOND PAIN REASSESSMENT]**

G. SECOND PAIN REASSESSMENT: Within 30 minutes (90 minutes after triage)

- Assess vitals
- Perform follow up lab tests* or review lab results and address abnormalities
- Re-evaluate for serious complications (see **D [TARGETED EVALUATION]**)

Assess pain using VAS or verbal scale (1-10):

If VAS ≥ 7 :

If no hypoxia or sedation:

- Repeat 2nd dose IV opioid (3rd dose) if route is available (if route is not available, consider other routes)
- Escalate 2nd dose of IV opioid by 25%

If signs of excessive sedation:

- Decrease dose of IV opioid

Consider adjunctive NSAIDs or acetaminophen (assess renal/liver function as needed):

If VAS 5-<7:

If no hypoxia or sedation:

- Repeat 2nd dose IV opioid (3rd dose) if route is available (if route is not available, consider other routes)
 - Escalate 2nd dose of IV opioid by 25%
 - Consider switching opioid*
- If signs of excessive sedation:
- Decrease dose of IV opioid

If VAS ≤ 4 :

- Offer short-acting oral opioid
- Assess if long-acting oral pain med prescribed as outpatient:
- If yes, restart long-acting oral pain med
 - If no, call for pain service consult or SCD provider team*

Ready for discharge (see **J**)

[DISCHARGE]

- Call hematology/SCD expert about patient being readied for discharge*

Consensus order set



I. PREVENTIVE CARE	
Consider vaccinations: <ul style="list-style-type: none"> <input type="checkbox"/> Consult CDC vaccination schedules^{vii} <input type="checkbox"/> If under age 5, twice-daily prophylactic penicillin* <input type="checkbox"/> Pneumovax (wait ≥8 weeks since prior Prevnar)* 	
Inquire about access to behavioral health/psychiatric services: <ul style="list-style-type: none"> <input type="checkbox"/> Order psychiatric referral* 	Consult Case Management and social work: <ul style="list-style-type: none"> <input type="checkbox"/> Support enrollment in appropriate services (e.g., disability)
J. DISCHARGE	
<ul style="list-style-type: none"> <input type="checkbox"/> Confirm patient's pain is adequately controlled <input type="checkbox"/> Schedule outpatient follow-up with PCP, hematology, or other SCD expert within 1 week 	
Determine the patient's current supply of pain medication	
If patient has adequate supply of pain medication, do not prescribe	If patient does not have adequate supply of pain medication: <ul style="list-style-type: none"> <input type="checkbox"/> Check prescription monitoring program (I-STOP)^{viii} <input type="checkbox"/> Prescribe 3-day supply of opioids.* Consider 5-7-day supply. <input type="checkbox"/> Prescribe adjunctive NSAIDS (consider renal function; should not be prescribed alone) <input type="checkbox"/> Prescribe constipation prophylaxis
Provide and review SCD Pain Home Management discharge instructions and SCD education:	
<ul style="list-style-type: none"> <input type="checkbox"/> Discuss signs of serious complications and instruct patient to return to ED if experience (e.g., acute chest syndrome, stroke, sepsis, fever, etc.) <input type="checkbox"/> Discuss addiction awareness <input type="checkbox"/> Discuss overdose signs <input type="checkbox"/> Prescribe Naloxone kits (for self and family members) if receiving ≥50 mg/day morphine equivalent dose <input type="checkbox"/> Consider recommending that the patient discusses other disease modifying treatments with hematologist: <ul style="list-style-type: none"> <input type="checkbox"/> Hydroxyurea <input type="checkbox"/> L-glutamine^{ix*} <input type="checkbox"/> Discuss setting up individualized treatment plan with SCD provider 	

Limitations



- The order set was developed by and for NYC clinicians and may not be generalizable to SCD care across the United States
- Whether this order set improves outcomes has not yet been demonstrated
- Despite high median ratings, some panelists disagreed on some items – the order set should be adapted to individual clinic settings
- Only 10 clinicians were involved, who brought their individual judgement and experience to the process

Conclusions



- A valid, reliable, and reproducible method was used to develop an order set to help standardize care for patients experiencing VOEs in NYC EDs
- Items in the order set have been shown to improve outcomes:
 - Implement or establish a patient’s SCD plan
 - Implement rapid triage (ESI level 2)
 - Initiate analgesic therapy within 30 minutes of triage
 - Assess pain repeatedly throughout the visit
 - Schedule follow-up appointments at discharge
- Consistent with the National Heart, Lung, and Blood Institute (NHLBI) Guidelines and Community Care of North Carolina Sickle Cell Task Force local protocol
- **Implementation of this order set in NYC EDs is ongoing**

References



1. Wang Y, et al. Genet Med. 2013;15(3):222-228
2. Brousseau DC, et al. JAMA. 2010;303(13):1288-1294
3. Kavanagh PL, et al. Pediatrics. 2015;136(4):e1016-e1025
4. Bemrich-Stolz CJ, et al. Int J Hematol Ther. 2015;1(1)
5. Tanabe P, et al. Acad Emerg Med. 2007;14(5):419-425
6. Lazio MP, et al. Clin J Pain. 2010;26(3):199-205
7. Tanabe P, et al. Acad Emerg Med. 2012;19(4):430-438
8. Ender KL, et al. Pediatr Blood Cancer. 2014;61(4):693-696
9. Community Care of North Carolina Sickle Cell Task Force. Emergency Department Vaso-occlusive Crisis Management: Adults and Children. 2018. <https://sickleemergency.duke.edu/sites/default/files/ccnc-voc-protocol.pdf>