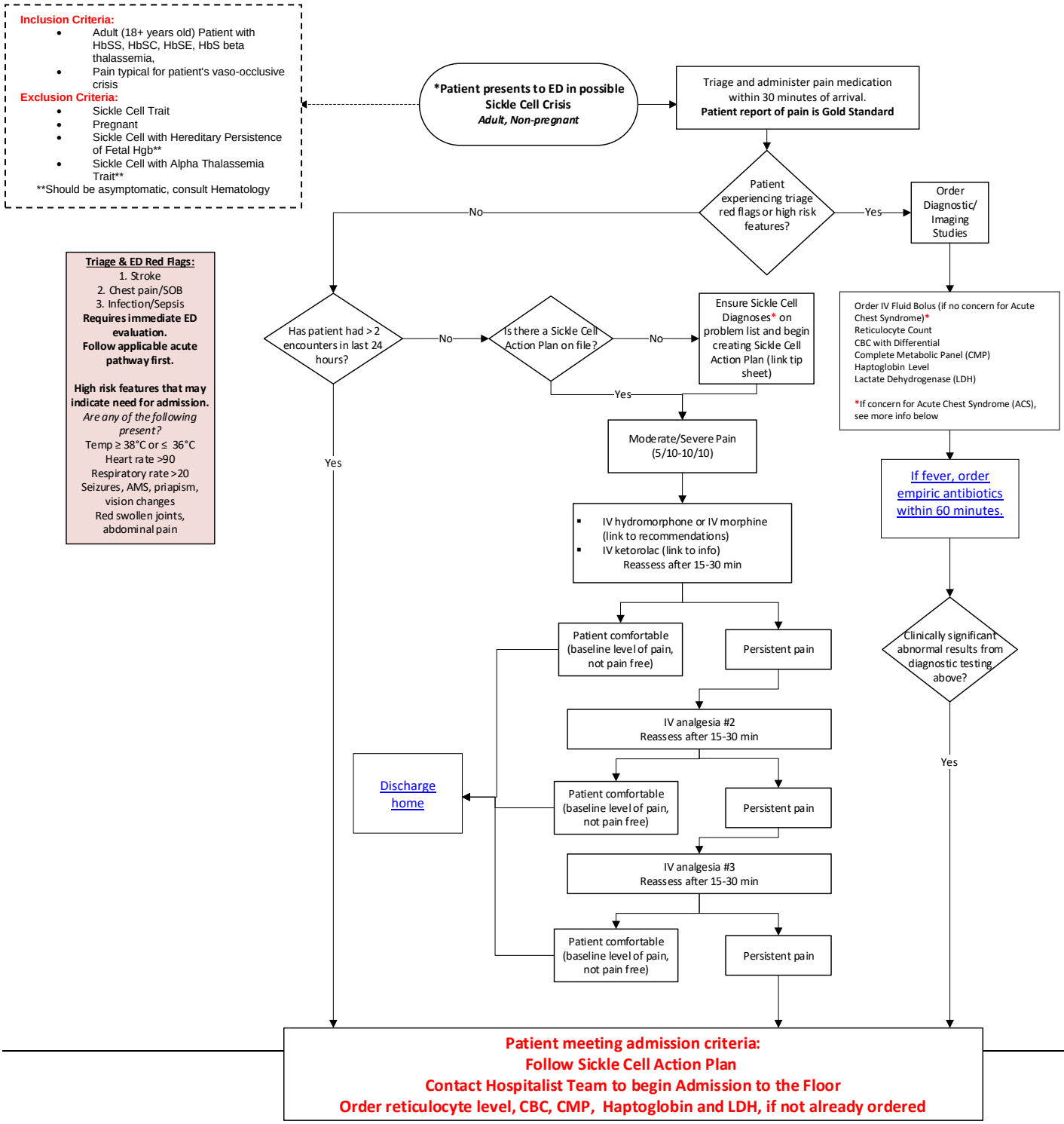
SICKLE CELL CRISIS, ADULT, INPATIENT AND EMERGENCY DEPARTMENT

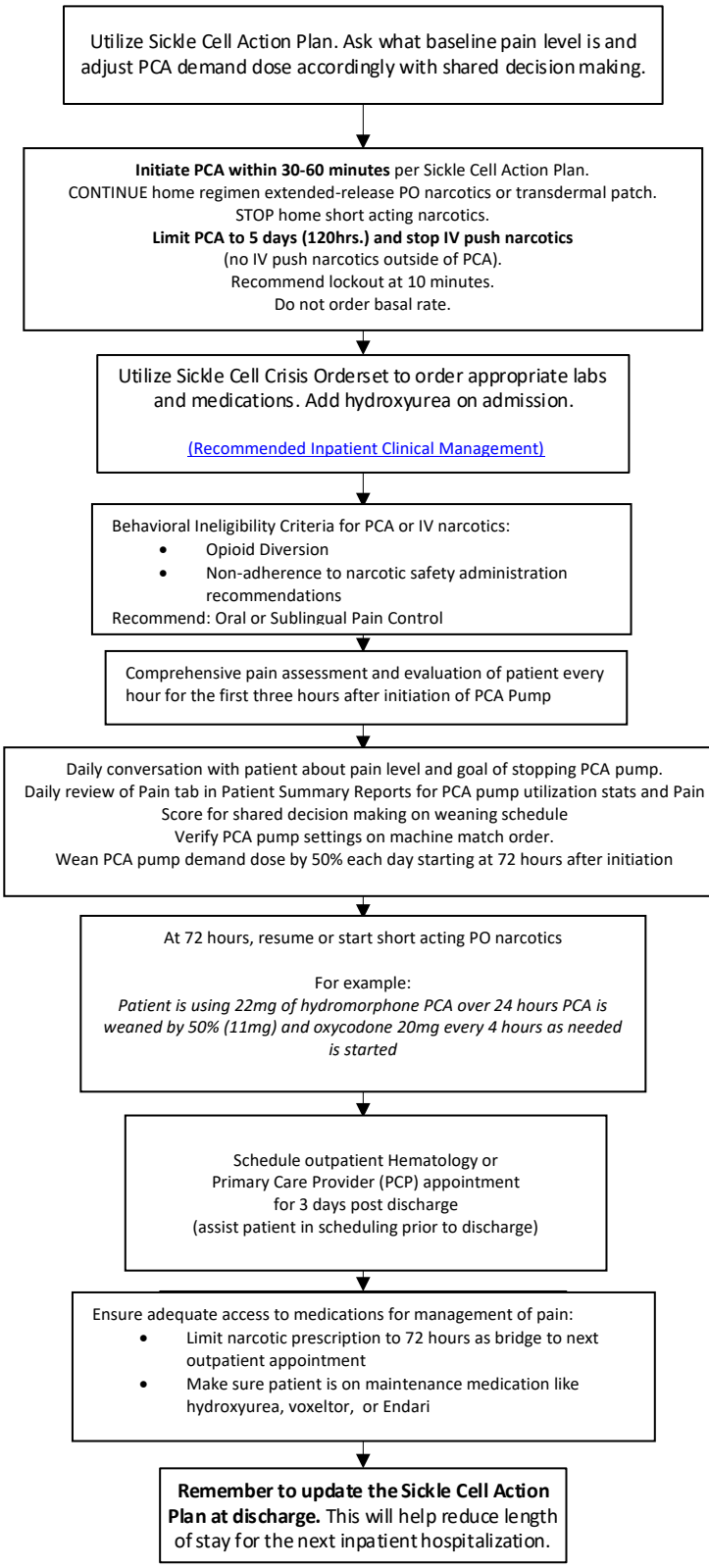
Updated: November 13, 2023

Emergency Department Clinical Algorithm:



Inpatient Clinical Algorithm:

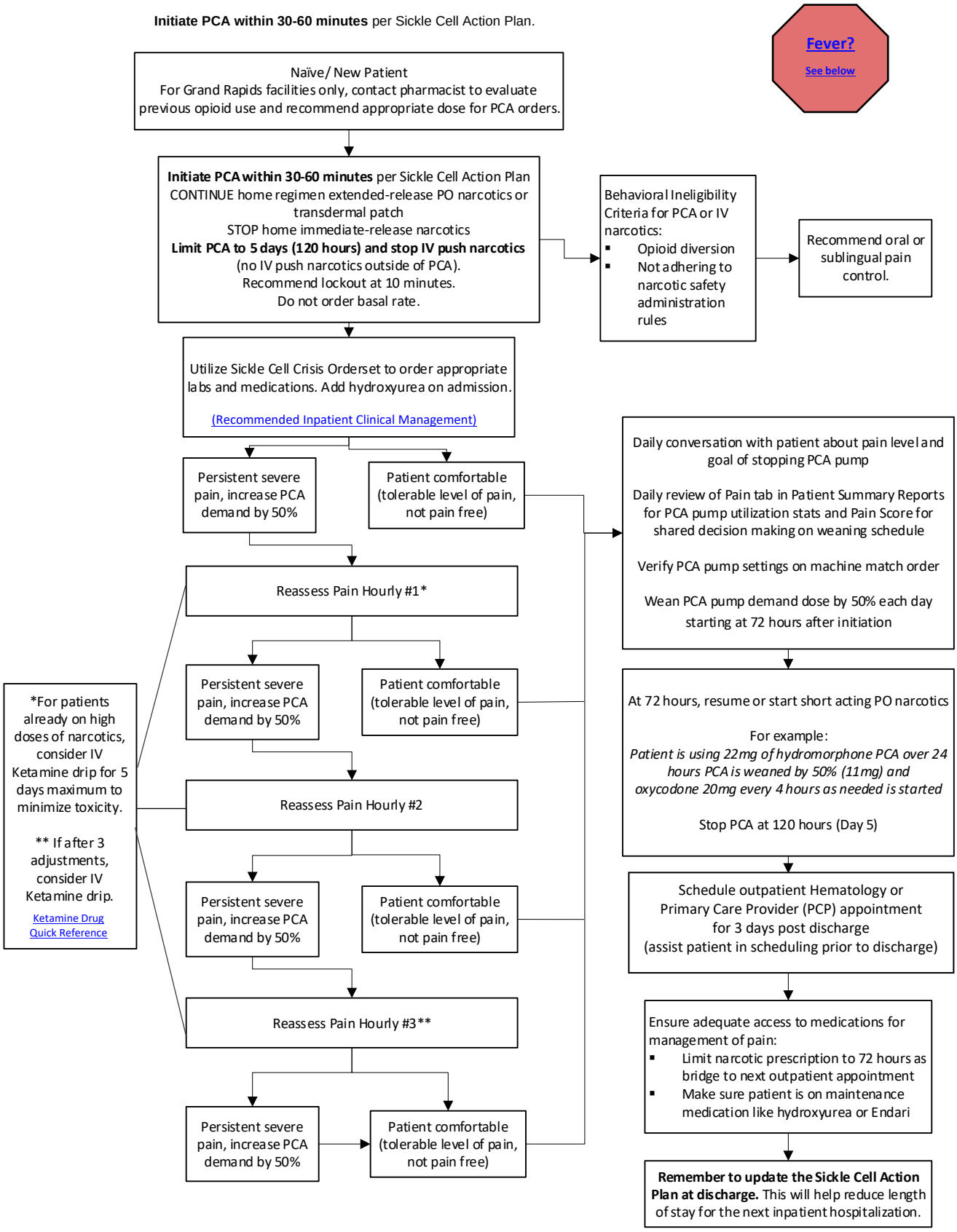
Established Patient or Patient with Verified Action Plan from outside institution**

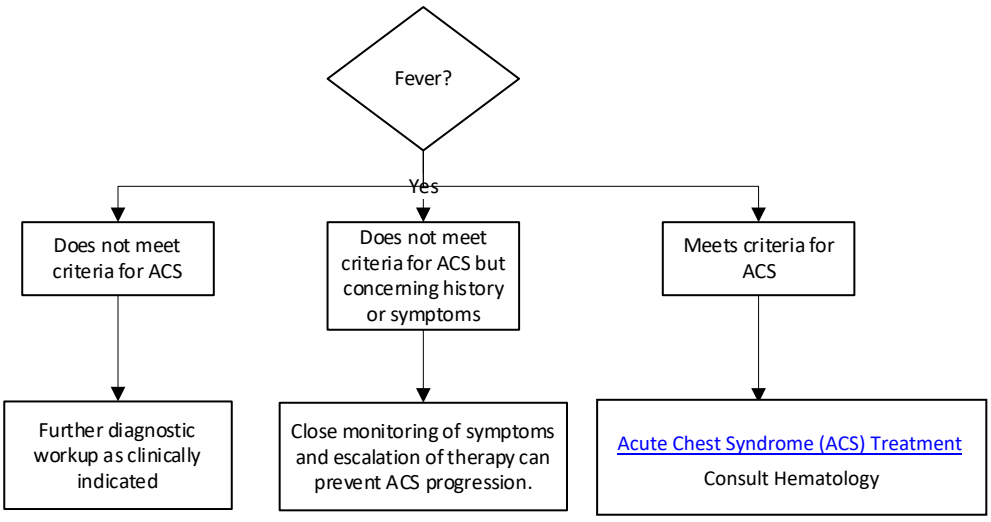


**Patients planning to establish care at Corewell Health with Sickle Cell Action Plan from outside institution that does not utilize PCA pump, consult pharmacy for conversion to PCA pump.

Inpatient Clinical Algorithm:

Naïve / New Patient





Clinical Pathway Summary

CLINICAL PATHWAY NAME: Sickle Cell, Adult, Inpatient and Emergency Department

PATIENT POPULATION AND DIAGNOSIS:
Inclusion Criteria:

- Adult (18+ years old) Patient with HbSS, HbSC, HbSE, HbS beta thalassemia not currently followed by Pediatric Hematology
- Pain typical for patient's vaso-occlusive crisis

Exclusion Criteria:

- Sickle Cell Trait
- Pregnancy
- Sickle Cell with Hereditary Persistence of Fetal Hgb**
- Sickle Cell with Alpha Thalassemia Trait**

APPLICABLE TO: Corewell Health West

BRIEF DESCRIPTION: How to manage Sickle Cell pain crisis for hospitalists and emergency medicine.

OPTIMIZED EPIC ELEMENTS:
Sickle Cell Orderset
Sickle Cell Action Plan

IMPLEMENTATION DATE: December 2023

LAST REVISED: 11/13/2023

Clinical Pathways Clinical Approach

ED TREATMENT AND MANAGEMENT: Sickle Cell | ACEP

Within 15-30 minutes of triage:
Begin administration of IV analgesia
Order 3 doses of IV analgesia PRN for moderate pain (5-7) to severe pain (rating 7-10)
Reassess every 15-30 minutes from dose administration and re-dose until pain improves

Administer according to Sickle Cell Action Plan on file or reference starting doses below for new patients.
Remember to note what medications were tried at home, doses taken, and time of last dose.

Recommended starting IV analgesia dosing for patients without Sickle Cell Action Plan:
IV Morphine 2-6mg x 3 doses
or
IV Hydromorphone 1-3mg x 3 doses

Recommended anti-inflammatory medication:
IV/PO Ketorolac*
*If no renal insufficiency or NSAID in last 6 hours and not on anticoagulation

AVOID IV diphenhydramine

Recommended Inpatient Clinical Management (once patient is admitted) in addition to PCA:

- Hydroxyurea 500 mg q/day if not already on a prophylactic sickle cell drug. Confirm patient is not pregnant prior to starting hydroxyurea. Patient may be a candidate for exchange transfusions outpatient if pregnancy planning.
    - Compliance with hydroxyurea is indicated by macrocytosis or elevated MCV unless they have concomitant thalassemia
- Incentive spirometry to prevent ACS
- Encourage early, progressive mobility
- DVT prophylaxis to prevent VTE
- D5 ½ normal saline or ½ normal saline limited to 24-48 hours. Avoid fluid bolus if ACS is suspected
- Folic acid 1mg daily
- Scheduled acetaminophen
- Scheduled ketorolac for 48 hours and then PRN for up to 5 days total (if no renal insufficiency or therapeutic anticoagulation or pregnant)
- Scheduled gabapentin
- Lidocaine patch PRN
- Fioricet for headache (or consideration for ketamine, especially for liver concerns because Fioricet contains acetaminophen)
- Avoid trazodone in men for sleep (causes painful priapism or erections in men with sickle cell anemia)
- Annual hemoglobin fractionation if not already done*
- **CBC with differential** and **reticulocyte** counts daily until clinical improvement
    - Rising reticulocyte count could indicate an impending drop in hemoglobin and worsening pain.
- PT/OT: encourage early, progressive mobility
- Social Work and or Clinical Psychology consult for mental health and/or psychosocial complications
- Care Management consult for discharge planning

***Hematology requests Hemoglobin (HgB) Fractionation be updated annually.**
If a patient is on exchange transfusions, Hematology will update the HgB Fractionation quarterly.
Remember, this tells us the percent of Hemoglobin S in the patient's blood and what type of sickle cell anemia that they have (the pathologist will addend the HgB Fractionation to indicate the sickle cell type, once resulted it can be found by scrolling to the very bottom of the lab).

Empiric Therapy of Fever (patients with SCD and fever are at risk for sepsis)
Treat with IV/IM ceftriaxone and azithromycin.
If cephalosporin allergy, use penicillin-based drug.
If cephalosporin and PCN allergy, use Levaquin.

Acute Chest Syndrome (ACS)
Definition: (cite up to date)
ANY new pulmonary density on chest imaging with at least one complete lung segment AND at least one of the following:

- Temperature ≥ 38.5°C
- 3 percent decrease in SpO2 from a documented steady-state value on room air
- Tachypnea
- Intercostal retractions, nasal flaring, use of sternocleidomastoid muscles in neck
- Chest pain
- Cough
- Wheezing
- Rales

Pneumonia can formally be considered as meeting the criteria for ACS (the two cannot be reliably distinguished)
A rapidly dropping platelet count can indicate impending decline in patient.
History of ACS puts one at higher risk for ACS in the future. It is important to document this in the sickle cell action plan.
Rapidly start empiric antibiotics within 60 minutes of identifying ACS. See Fever box for antibiotics.
If concern for ACS, no IV Fluid Bolus but run maintenance fluids at slow rate. (Reference Johns Hopkins)

Uptodate describes a need for a high level of vigilance as delays in diagnosis can adversely impact outcomes. Prompt initiation of exchange transfusions could be lifesaving.

ACS Treatment (within 60 min of identification): (take a look at Johns Hopkins)
Additional laboratory studies

- Monitoring
- Respiratory support
    - If Oxygen is less than 92%, add oxygen, spirometry, airway clearance
    - Albuterol
- Fluid management (avoid IV fluid bolus)
- Antibiotic recommendations
- Analgesia
- Transfusion therapy
- Hematology Consultation for emergent exchange transfusion, ICU considerations

Discharge with resources/condition specific instructions and on maintenance medications:

- Hydroxyurea (PO), crizalizumab-tmca, L-glutamine, Endari, or Voxelotor
- Patient follow up appointment with a hematologist/PCP
- Consider Consult with Hematology for Discharge Recommendations
- Review PDMP
- Limit narcotic prescription to 72 hours and advise patient to follow up with primary prescriber
- GOAL = Baseline level of pain, NOT Pain free
- Document if narcotic prescribed in Sickle Cell Action Plan

Things to remember:

- Patients with Sickle Cell Disease are prioritized for hospital units with specialty trained nurses and pharmacists: CHW Blodgett 4E and 3D.
- Remember to address other comorbidities like hypertension, arthritis, etc. when a patient is hospitalized.
- Be vigilant in identifying other causes of pain that may need additional treatment. SCD complications can be elusive due to the failure to look beyond the underlying pathology during painful sickle cell crisis.
- Discuss possible triggers of crisis with patient: Infection? Cold and windy weather? Overexertion from a recent apartment move? Stress from social issues? Alcohol use? Involve MSW if appropriate.
- The objective of PCA is to break the crisis safely and quickly. The goal is to transition to oral medications to provide sustained pain relief and transition to an oral regime patient can discharge with. Pain may not fully be controlled by Day 5 and pharmacy will help determine oral equivalent of PCA dosing.
    - Before adjusting PCA, review the pain flowsheet and see how many demand doses the patient has received. It may be that the patient is not pushing the button enough.
    - If you adjust the demand dose, make sure you also adjust the 4 hour lock out.

- In your progress note, indicate if you are okay with the APP or remote physician adjusting PCA. Excess use of IV pushes sometimes means that the patient is not pushing the demand dose. Encourage the patient to push the demand dose button if they are still in pain so that we can use this information to adjust the PCA. PCA data can be found under “PAIN” in flowsheets.
- Be vigilant in identifying other causes of pain that may need additional treatment. SCD complications can be elusive due to the failure to look beyond the underlying pathology during painful sickle cell crisis.
- Make sure patients are on medications like hydroxyurea (to reduce or prevent multiple complications), crizalizumab-tmca (to reduce vaso-occulsive crisis), voxelotor (to prevent the sickling of red blood cells). L-glutamine (to reduce pain crises) etc. Consult hematology if you need help. Encourage patients to take medications to help reduce the chances of acute pain crises, hospitalizations, and other complications. Patients chronically in the hospital who are unable to follow up or establish care with hematology, request an inpatient hematology consult to help break this cycle.
- Let’s change our vocabulary: Use of the term “sickler” is considered derogatory and offensive as it reduces the patient to a disease instead of being viewed as a patient in need.
- 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

Pathway Information

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EXPERT IMPROVEMENT TEAM (EIT): Sickle Cell EIT

CLINICAL PRACTICE COUNCIL (CPC): Acute Health

CPC APPROVAL DATE: 12/5/2023

OTHER TEAM(S) IMPACTED: Nursing, Hematology, Hospitalists, Emergency Department

References

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Acute chest syndrome (ACS) in sickle cell disease (adults and children) - UpToDate

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