Sickle Cell Pain Crisis

Aliases

None

Patient Care Goals

1. Identify potentially life-threatening complications of a sickle cell disease.
2. Improve patient comfort.

Patient Presentation

Inclusion Criteria

1. Patient with known sickle cell disease experiencing a pain crisis

Exclusion Criteria

1. Pain due to acute traumatic injury [see Trauma section guidelines]
2. Abdominal pain due to or related to pregnancy [see OB/GYN section guidelines]
3. Patients with sickle cell trait

Patient Management

Assessment

1. Perform airway assessment and management per the Airway Management guideline.
2. Obtain vital signs (including pulse, respiratory rate), pulse oximetry, and blood pressure
3. Provide evaluation and management of altered mental status per the Altered Mental Status guideline.
4. Provide evaluation and management of pain per the Pain Management guideline.
5. Obtain vascular access as necessary to provide analgesia and/or fluid resuscitation.
6. Assess for potentially serious complications other than pain crisis which may include:
    a. Acute chest syndrome
        i. Hypoxia
        ii. Chest pain
        iii. Fever
    b. Stroke [see Suspected Stroke/Transient Ischemic Attack guideline]
        i. Focal neurologic deficits
    c. Meningitis
        i. Headache
        ii. Altered mental status
        iii. Fever
    d. Septic arthritis
        i. Severe pain in a single joint
        ii. Fever
    e. Splenic sequestration crisis (usually young pediatric patients)
        i. Abdominal pain, LUQ
        ii. Splenic enlargement (examine with care)
        iii. Hypotension, tachycardia
    f. Severe anemia
        i. Pallor
        ii. Fatigue
        iii. Dyspnea or dyspnea on exertion
        iv. Shock
    g. Infections: Pneumonia (cough, fever, sputum shortness of breath)
    h. Priapism: Painful, prolonged erection in the absence of sexual activity
    i. Venous thromboembolism; Calf pain, tenderness, swelling, chest/back pain especially with inspiration, shortness of breath

7. Assess for signs of shock—if shock is present, treat per Shock guideline

Treatment and Interventions

1. Medication Administration:
    a. Provide analgesia per the Pain Management guideline.
    b. Administer oxygen as appropriate for dyspnea or distress with a target of achieving 94-98% saturation for most acutely ill patients
    c. Start an IV and provide saline 10ml/kg normal saline bolus (up to 1L) ***[AEMT].***
    d. Provide transport to an appropriate receiving facility.
    e. Reassess vital signs and response to therapeutic interventions throughout transport.
2. Comfort measures:
    a. Keep patient warm and dry.
    b. Transport in a position of comfort unless clinical condition requires otherwise.

Patient Safety Considerations

None recommended

Notes and Educational Pearls Key Considerations

- Assess for life-threatening complications of sickle cell disease—these patients have significantly higher risk of numerous complications in addition to pain crises.
- Provide appropriate treatment for pain, respiratory distress, and shock.
- These patients may have a higher tolerance to narcotic pain medications if they are taking them on a regular basis.
- These patients will tolerate acute blood loss poorly due to baseline anemia.
- Patients with sickle cell trait can have acute pain crises in extreme conditions (e.g. heat exhaustion, dehydration) and a number of college athlete deaths have been linked to sickle cell trait.

Pertinent Assessment Findings

- Lung exam and assessment of respiratory distress
- Altered mental status
- Focal neurologic deficits
- Inability to move a joint

Quality Improvement

Associated NEMSIS Protocol(s) (eProtocol.01)

- 9914165—Other (as of 3/1/2017, no specific NEMSIS protocol exists)

Key Documentation Elements

- Documentation of normal respiratory and neuro status
- Documentation of how this pain crisis compares with others in terms of location, severity, and triggers
- Documentation of home pain medications used

Performance Measures

- Assessment for life-threatening etiology
- Mitigation of pain per the Pain Management guideline

Reference

1. Mitchell BL. Sickle cell trait and sudden death – bringing it home. *J Natl Med*

Assoc. 2007;99(3):300-5.