

SICKLE CELL CRISIS

(Last Updated: 08/12/2019; Reviewed by: Bibek Karki, MBBS)

PRESENTING COMPLAINT: Sudden onset of pain (over extremities, chest, back and abdomen), fever, respiratory distress, cough

FINDINGS

- **A** Check airway
- **B** Normal / ↑ RR, respiratory distress, ↓ SpO₂
- **C** Normal / ↑HR
- **D** Variable altered (VUPD), pain, seizure/focal deficit
- **E** Pain and swelling over the extremities
- **L_{PC}** ↓ Hb, ↓ platelet count, ↑ circulating nucleated RBCs
- **U_{PC}** Enlarged Spleen or liver. Rule out cholecystitis, cholelithiasis, nephrolithiasis or ectopic pregnancy, B lines in acute chest syndrome (ARDS)

***V** (verbal), **P** (pain), **U** (unconsciousness), **D** (delirious)

Upc (point of care ultrasound) **Lpc** (point of care labs)

OTHER HISTORY

- History of sickle cell disease, dactylitis (associated pain and swelling in hands and feet), priapism (painful and persistent erection of penis)
- Triggers: infection, cold exposure, fever, dehydration, acidosis, hypoventilation, low humidity, wind, stress

DIFFERENTIAL DIAGNOSIS

- Pneumonia, pulmonary emboli, bone marrow infarction and embolism, myocardial infarction

OTHER INVESTIGATIONS

- Monitor pain assessment and signs of fever
- Labs: CBC, PLT, reticulocyte count, blood culture, if febrile, LFT, Amylase/lipase for RUQ, epigastric, or severe abdominal pain
- Imaging
 - CXR (a new radiodensity)
 - Renal and liver function tests, if no prior evaluation
- Type and cross match, if Hgb is > 2 g below baseline

THERAPEUTIC INTERVENTIONS

- **Oxygen**

- Administer oxygen if patient has oxygen saturation < 95% on room air: give 2L/minute via nasal cannula
- Incentive spirometry: should use spirometer q2h while awake, decreases risk of atelectasis due to pain and decreases progression to acute chest syndrome
- Non-invasive ventilation: CPAP and BiPAP, for patients with poor respiratory effort
- Mechanical ventilation, for patients with respiratory failure and ARDS
- ECMO, if mechanical ventilation fails
- **Bronchodilator (inhaled):** For patients with history of reactive airway disease (asthma), even when the patient is not currently wheezing; and without history of prior reactive airway disease
- **Hydration**
 - 10-20 cc/kg followed by 1-1.5x's maintenance fluids with appropriate IV fluid
 - If hypovolemic, recommend 0.9% NS
 - If euvolemic, recommend 0.45% NS, since hypernatremia can precipitate sickling of RBC
- **Pain:** Analgesia should be selected based on pain assessment, associated symptoms, outpatient analgesic use, patient's knowledge of effective agents and doses, past experience with side effects
 - Consider NSAIDs for mild-moderate pain (ketorolac)
 - Opioids for severe pain; titrate based on pain severity; IV drugs, in order of choice: morphine, dilaudid, fentanyl; Meperidine is not preferred due to toxic metabolites: lowers seizure threshold and accumulates with renal insufficiency
- **Adjunctive therapy for analgesic side effects:** Antihistamine, such as diphenhydramine, to offset histamine released by mast cells due to opioids
- **Antibiotics:** Used as a prophylactic measure; broad spectrum antibiotic coverage should be immediately started for all patients with ACS
 - Third generation cephalosporin + Macrolide
 - Clindamycin + Macrolide, if allergic to cephalosporin
 - Third generation cephalosporin + Macrolide + Vancomycin, for severely ill patient with pulmonary infiltrate
- **Transfusion:** Also improves oxygenation
- **Acetaminophen:** If fever is present

ONGOING TREATMENT

- **Prophylaxis**
 - **Bowel Regimen:** Used to reduce opioid-induced constipation
 - Docusate to soften stool, Senna to induce bowel motility
 - If no bowel movement with docusate and Senna: consider increasing dose
 - If no bowel movements by day 4-5: add bisacodyl or enema

- **Hydroxyurea:** Used if ≥ 3 crises in the past 12 months or interferences with daily activity, decreases crises by an average of 50%, decreases risk of acute chest syndrome
 - Initiate at a low dose and gradually increased to a dose that does not cause severe hematologic toxicity
 - Monitor CBC
- **Transfusion:** For those who continue to have ACS episodes despite hydroxyurea therapy
- **Vaccination:** All routine vaccination, especially pneumococcal vaccination
- Close follow up with Hematologist

CAUTIONS

- Monitor for acute chest syndrome for possible exchange transfusion
- Antibiotics: used in acute chest syndrome and cases of fever and infection
- Transfusion indications:
 - Simple transfusion: Symptomatic acute chest syndrome (ACS) and decreased hemoglobin 1 g/dL below baseline; acute splenic sequestration plus severe anemia; aplastic crisis; symptomatic anemia
 - Exchange transfusion: Symptomatic severe ACS (oxygen saturation $< 90\%$, despite supplemental oxygen)
 - Simple or exchange transfusion: Stroke, hepatic sequestrations, intrahepatic cholestasis, multisystem organ failure with exchange or simple transfusion
 - Transfusion is not indicated for uncomplicated painful crisis, priapism, asymptomatic anemia, acute kidney injury (unless multisystem organ failure)

REFERENCES & ACKNOWLEDGMENTS

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