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
- LPC ↓ Hb, ↓ platelet count, ↑ circulating nucleated RBCs
- UPC 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
o Administer oxygen if patient has oxygen saturation < 95% on room air: give 2L/minute via nasal cannula
o Incentive spirometry: should use spirometer q2h while awake, decreases risk of atelectasis due to pain and decreases progression to acute chest syndrome
o Non-invasive ventilation: CPAP and BiPAP, for patients with poor respiratory effort
o Mechanical ventilation, for patients with respiratory failure and ARDS
o 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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