Acute Chest Syndrome in Sickle Cell Disease Care Guideline

Inclusion Criteria:
- Children with sickle cell disease
- New pulmonary infiltrate on chest x-ray, involving at least one complete lung segment, excluding atelectasis

Assessment
- Admit to 5S or PICU
- NPO
- History and physical with focus on pulmonary exam
- Vitals, accurate height and weight
- Strict I/O; Daily Weight
- O2 saturation
- Diagnostics: CBC with Retic, CMP with LDH, STAT Type and Screen, Hgb Electrophoresis
- Chest X-ray (CXR) – 2 View
- For significant respiratory distress, consider VBG
- Fever >38°C or elevated WBC, obtain Blood Culture
- URI symptoms, obtain Respiratory Panel PCR

Interventions
- Stat Hematology consult (if not admitted to Hematology service)
- Pulmonary consult
- Notify apheresis team as soon as possible during working hours if anticipated procedure
- Consult PICU for possible apheresis catheter placement
- Oxygen to keep O2 sat ≥94%
- Maintenance IV fluids
- Antibiotics: cefTRIAXone 75 mg/kg IV q24h (max dose 2g) and azithromycin 10 mg/kg IV q24h (max dose 500mg)
- Pain medication: Ketorolac 0.5 mg/kg IV q6h (max dose 30mg) for 5 DAYS PRN Pain (Moderate)
- Cardiac monitoring
- Simple transfusion of PRBCs to **max** Hgb = 10 gm/dl
- Repeat CXR, CBC STAT if worsening respiratory symptoms/increased oxygen requirements
- Consider exchange transfusion (see order set) if:
  - initial Hgb ≥ 10 gm/dl
  - worsening respiratory symptoms
  - worsening radiological findings (CXR), despite simple transfusion and supportive care
- Albuterol HFA MDI 2 PUFFS Q4hr PRN wheezing or poor air exchange; Chest Physiotherapy after each treatment - Use Vest if patient > 10 kg (See Policy RT205)
- Incentive Spirometry (IS) for patients ≥ 5years, every 2 hrs while awake - use bubbles or pinwheel if patient < 5 years or unable to do IS

Further Recommendations
- Consider furosemide for s/s of fluid overload
- Consider systemic steroid if wheezing
- Cardiology evaluation with Echo and EKG
- Keep Hgb > 9 gm/dl for 2-3 months post discharge

Discharge Criteria
- Afebrile > 24 hours
- Baseline oxygen requirement; improved respiratory symptoms
- On oral antibiotics
- Stable Hgb
- Adequate pain control on oral medication
- Adequate oral intake
- Catch up vaccines, specifically Pneumovax 23 (every 5 yrs); seasonal influenza; COVID-19 (recommended, if age appropriate)

Recommendations/ Considerations

Predictors:
- Pain crisis involving chest, shoulders and back
- Post anesthesia complication
- Respiratory Infections
- On narcotics
- Asthma exacerbation

- Baseline Hgb level may run low < 9gm/dl

- If suspected pulmonary embolism, obtain CT angiogram of chest

- May need more than 1 exchange transfusion if clinical findings not improving

- After recovery from acute crisis, patient should be started on hydroxyurea if not already taking; optimize dose

- History of more than 1 acute chest crisis, consider chronic transfusion protocols to keep HgbS < 25%

- If recurrent crises, consider BMT if match available

- Monitor for right ventricular (RV) failure and/or pulmonary hypertension

- If continued worsening despite above interventions, will need respiratory support, including non-invasive ventilation, intubation, iNO, or ECMO

- Apheresis team hours 7a-7p weekdays, 7a-7p weekends (see call schedule on PAWS)

Patient/Parent or Caregiver Education
- Asthma education/AAP if has evidence of asthma

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References

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