Acute Chest Syndrome in Sickle Cell Disease Care Guideline

CHOC Children's

Inclusion Criteria:
- Children with sickle cell disease
- New pulmonary infiltrate on CXR, 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
- Consider VBG for significant respiratory distress
- CXR 2 view
- Blood culture if fever >38, elevated WBC
- VRP if URI symptoms

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 and azithromycin
- Pain medication: toradol
- 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 Q4hr, Chest PT after each treatment. Use Vest if pt > 10 kg
- Incentive Spirometry for pts >= 5yrs, every 2 hrs while awake - use bubbles or pinwheel if pt < 5 yrs 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 d/c

Discharge Criteria
- Afebrile > 24 hrs
- 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 (every 5 yrs); seasonal influenza

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, pt 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 RV 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, 8a-8p weekends (see call schedule on PAWS)

Patient/Parent or Caregiver Education
- Catch up vaccines, specifically pneumovax (every 5 yrs); seasonal influenza
- Asthma education/AAP if has evidence of asthma

Approved by Evidence Based Medicine Committee 11/16/2016

References
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