

Children's Mercy Kansas City

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Care Process Models

Quality Improvement and Clinical Safety

1-2021

Acute Chest Syndrome in the Sickle Cell Patient

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These guidelines do not establish a standard of care to be followed in every case. It is recognized that each case is different and those individuals involved in providing health care are expected to use their judgment in determining what is in the best interests of the patient based on the circumstances existing at the time. It is impossible to anticipate all possible situations that may exist and to prepare guidelines for each. Accordingly, these guidelines should guide care with the understanding that departures from them may be required at times.

Patient with Sickle Cell Disease experiencing Acute Chest Syndrome (ACS) in the ED

Algorithm finalized/ revised: 1.4.2021
Owner: Kayeleigh Higgerson, DO, Fellow, PGY-5
Mentor: N. Tabassum Iqbal, MD

Obtain history of:

- Prior ACS
- Asthma
- Respiratory symptoms (cough, shortness of breath, dyspnea)
- Fever
- Current chest pain
- Restrictive lung disease
- Nocturnal hypoxia
- Recent history of sedation w/o prior transfusion

If patient is known to CM: Review Critical Information note

Initial pt. work-up to include:

- CBC w/differential, reticulocyte count, HbS level
- BMP, liver function panel, LDH
- Consider blood gas
- Blood culture if febrile, hypotensive or toxic-appearing
- Consider RVP and COVID-19 testing
- Type and Screen
- Chest X-Ray (2 views) for any respiratory symptoms, even in absence of hypoxia or abnormal lung findings on exam

Management of patient while in ED and transferred to either Hem/Onc resident service or PICU (dependent on pt. status):

- Consult Hem/Onc
- Oxygenation:
 - Supplemental oxygen only if hypoxic (O₂ saturation: <94% or >4% below baseline if known chronic hypoxia)
 - Incentive spirometry q 2 hours with Respiratory Therapy while awake using age appropriate respiratory therapy (pinwheel and bubbles); encourage pt. to accomplish hourly
 - IPPB q 4 hours as indicated
 - Consider trial of bronchodilators (atrovent/albuterol) in pts with history of asthma or wheezing on exam. Follow asthma action plan in pts with history of asthma.
- Encourage ambulation and activity
- Pain Management:
 - PO tylenol and NSAIDS
 - IV narcotic, refer to critical note for preferred narcotic
- Fluid Management:
 - Make NPO
 - Avoid fluid bolus as this may exacerbate pulmonary edema
 - Start IV fluids at three quarters to full maintenance rate
- Empiric antibiotics (Ceftriaxone and Azithromycin)
 - If pt. allergic to cephalosporin then consider Clindamycin
- Simple transfusion if Hgb > 2 gm/dL below baseline
 - Transfuse packed red blood cells (discuss volume with Hem/Onc consultant)
 - Sickle negative and cross-matched for C, E, Kell antigens*
 - Goal: Hgb 10 - 11 gm/dL
 - Labs after transfusion: CBC, retic, and HbS

Is the pt. exhibiting signs necessitating PICU care?

• Transfer/Continue care in PICU
• Consult Hem/Onc Service
• Prepare for exchange transfusion

Is pt. stable for transfer to floor?

Transfer to Hem/Onc Service

Management per Hem/Onc and Discharge Home

References used to establish this care standard

