1-2021

Acute Chest Syndrome in the Sickle Cell Patient

Children's Mercy Kansas City

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

Algorithm finalized/revised: 1.4.2021
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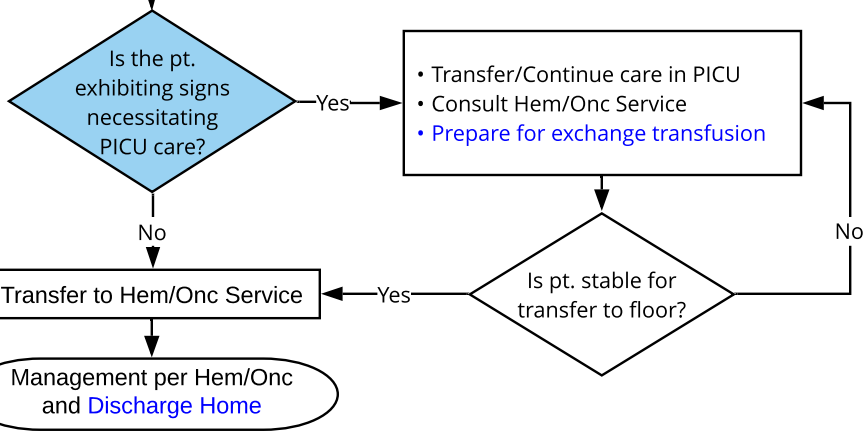
Obtain history of:
- Prior ACS
- Asthma
- Respiratory symptoms (cough, shortness of breath, dyspnea)
- Fever
- Current chest pain
- Restrictive lung disease
- Noctural hypoxia
- Recent history of sedation w/o prior transfusion

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 (O_2 saturatation: <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



References used to establish this care standard