Sickle Cell Disease (Pediatric) Order Set

Reason / Problem UpToDate

- *For patients with sickle cell disease (SCD), acute chest syndrome (ACS) is the second most common cause of hospitalization (second to vasoocclusive pain). It is the most common cause of death, with one-fourth of SCD-related deaths due to ACS.*
- *The diagnosis of acute chest syndrome requires a new pulmonary infiltrate on chest radiography that involves at least one complete lung segment, and one or more of the following: chest pain; temperature greater than 38.5ºC; hypoxemia; and signs of tachypnea, wheezing, cough, or the appearance of increased work of breathing.*

Sickle Cell Anemia UpToDate UpToDate UpToDate

Diet

☐ Nothing by mouth

Vital Signs

☐ Check vital signs per protocol [ ]

☐ Check vital signs [ ] [ ] [ ] [ ]

IV UpToDate

- *If dehydration is present, it should be corrected, as hypovolemia can contribute to increased sickling. Hypovolemia should be corrected with the administration of isotonic solution.*
- *Overhydration or rapid hydration should be avoided because they may result in pulmonary edema or heart failure.*

Crystalloid:

☐ 5% dextrose in quarter-normal saline [ ] [ ] [ ] [ ]

☐ Normal saline in peripheral venous line [ ] and [ ] [ ] [ ]

Crystalloid-Fluid Bolus:

☐ Normal saline bolus [ ] [ ] [ ]

Lock IV:

☐ Saline lock IV [ ] [ ]

Blood Products: UpToDate

- *In patients with acute chest syndrome (ACS), transfusion therapy improves oxygenation and should be considered early in the management of these patients.*

☐ Infuse packed red blood cells, crossmatched [ ] [ ] [ ]

Other Nursing

- *Because fever may be the first and only indication of a serious bacterial infection in a patient with sickle cell disease (SCD), patients and/or parents should be counseled to seek prompt medical attention for a predetermined elevated temperature. This should occur even if the fever rapidly goes away with or without antipyretics. (UpToDate)*

Assessments:

☐ Complete pediatric pain assessment [ ]

☐ Obtain weight in kilograms [ ]

☐ Perform admission assessment pediatric

Cardiac:

☐ Continuous bedside cardiac monitoring [ ]

Circulatory:

☐ Peripheral IV line care per protocol

☐ Insert peripheral IV line [ ]

☐ Central venous line care per protocol

Education:

- *Education of the child's parents or caregivers about the routine and emergency complications of sickle cell disease (SCD) is a cornerstone of effective medical care.*

☐ Provide disease/medical condition education [ ] UpToDate

Fluid Balance:

☐ Fluid volume management per protocol

☐ Intake and output [ ] and then [ ] [ ]

Respiratory:

- *Provide supplemental oxygen to maintain the oxygen saturation at patient's baseline value, generally above 90 percent. (UpToDate)*

☐ Evaluate breath sounds

☐ Maintain oxygen saturation greater than or equal to 90%

☐ Monitor pulse oximetry continuously, notify MD if oxygen saturation is less than 90%

Urinary:

☐ Catheter care per protocol

☐ Insert Foley catheter [ ] [ ]

Therapies

Procedures:

☐ [ ] central venous catheter [ ]

☐ Lumbar puncture [ ] *Cerebrospinal fluid is obtained for analysis and culture only if there is a clinical suspicion for meningitis. (UpToDate)*

Respiratory Therapy Service:

- *Respiratory support is provided to maintain oxygen saturation greater than or equal to 92 percent and includes supplemental oxygen, frequent incentive spirometry, and continuous positive pressure ventilation devices. (UpToDate)*

☐ Endotracheal intubation per protocol [ ] [ ]

☐ Incentive spirometry [ ] [ ]

☐ Non-invasive ventilation

- [ ]
- [ ]
- [ ]
- [ ]
- [ ]
- [ ]

☐ [ ], [ ] oxygen [ ] [ ] titrate to keep Sp02 greater than or equal to 92%

☐ Ventilator management [ ]

Medications

- *After the initial laboratory tests are obtained, UpToDate recommends prompt administration of empiric parenteral antibiotics in all patients with sickle cell disease (SCD) and fever (Grade 1C). (UpToDate)*

Anesthetics:

☐ Lidocaine HCl 1 % solution 0.25 mL subcutaneously via needle-free injection system (J-Tip) single dose prior to IV insertion as needed for anesthesia

Antibiotics: UpToDate

- *Because infection is one of the most common causes of acute chest syndrome (ACS), UpToDate recommends broad spectrum antibiotic coverage for patients presenting with ACS (Grade 1B). UpToDate typically uses a third generation cephalosporin (eg, cefotaxime or ceftriaxone) for bacterial coverage, and a macrolide (eg, azithromycin or erythromycin) for coverage of atypical organisms.*
- *For patients who are allergic to cephalosporins, clindamycin can be used (dose of 10 to 15 mg/kg, maximum dose 1.6 g).*
- *For the severely ill patient, with large or progressive pulmonary infiltrates, consider adding vancomycin to cover bacteria that are resistant to cephalosporins, such as methicillin resistant Staphylococcus aureus (MRSA).*

☐ Azithromycin 10 mg/kg intravenously single dose (not to exceed 500 mg in 24 hours)

☐ Cefotaxime sodium 50 mg/kg intravenously single dose (not to exceed 2 grams per dose)

☐ cefTRIAXone sodium 50 mg/kg intravenously single dose (not to exceed 2 grams in 24 hours)

☐ Clindamycin phosphate 10 mg/kg intravenously single dose (not to exceed 4.8 grams in 24 hours)

☐ Erythromycin lactobionate 5 mg/kg intravenously single dose (not to exceed 4 grams in 24 hours)

☐ Vancomycin HCl 15 mg/kg intravenously single dose (not to exceed 1 gram per dose)

Antipyretics-Analgesics: UpToDate

- *Adequate analgesia of spine, thoracic, and abdominal pain is important to prevent hypoventilation. UpToDate suggests the initial use of Ketorolac as a nonsedating analgesic (Grade 2C).*
- *When using ketorolac, one should not simultaneously use other nonsteroidal anti-inflammatory agents (NSAIDs). (UpToDate)*

☐ Ketorolac tromethamine 0.5 mg/kg intravenously single dose (not to exceed 30 mg per dose)

☐ Acetaminophen 160 mg/5 mL suspension 15 mg/kg orally single dose (not to exceed 5 doses in 24 hours)

☐ Acetaminophen 650 mg orally single dose (not to exceed 5 doses in 24 hours)

☐ Acetaminophen 80 mg suppository 15 mg/kg rectally single dose (not to exceed 5 doses in 24 hours)

☐ Ibuprofen 100 mg/5 mL suspension 10 mg/kg orally single dose (not to exceed 600 mg per dose)

☐ Ibuprofen 400 mg orally single dose (not to exceed 600 mg per dose)

Corticosteroids: UpToDate

- *A brief course of corticosteroids may be used to treat a comorbid flare of asthma, but these should be tapered before they are discontinued to reduce the risk of rebound vasoocclusive crisis.*

☐ methylPREDNISolone sodium succinate 2 mg/kg intravenously single dose (not to exceed 60 mg in 24 hours)

☐ prednisoLONE 15 mg/5 mL syrup 2 mg/kg orally single dose (not to exceed 60 mg in 24 hours)

☐ predniSONE 2 mg/kg orally single dose (not to exceed 60 mg in 24 hours)

Diuretics:

- *Overhydration or rapid hydration should be avoided because they may result in pulmonary edema or heart failure. Furosemide may be helpful if fluid overload is suspected.*

☐ Furosemide 0.5 mg/kg intravenously single dose (not to exceed 6 mg/kg total dose)

Bronchodilators: UpToDate

- *UpToDate recommends the use of bronchodilators in patients with wheezing or a prior history of asthma (Grade 1B).*

☐ Albuterol sulfate 90 mcg/actuation aerosol 2 inhalations via metered-dose inhaler single dose

☐ Albuterol sulfate 2.5 mg/3 mL nebulizer solution 2.5 mg nebulized single dose (not to exceed 5 mg per dose)

☐ Albuterol-ipratropium 2.5-0.5 mg/3 mL nebulizer solution 3 mL nebulized single dose (not to exceed 3 doses per episode)

☐ Albuterol-ipratropium 2.5-0.5 mg/3 mL nebulizer solution 3 mL nebulized every 20 minutes for 3 doses

Opioid Analgesics:

- *For patients requiring morphine or other opioids, careful attention to dosing is required, as doses of morphine that are high enough to cause hypoventilation may cause atelectasis. This in turn may lead to ventilation-perfusion mismatch and intrapulmonary sickling.*
- *The use of patient controlled analgesia (PCA) may minimize oversedation and hypoventilation but still provide adequate pain control.*

☐ HYDROmorphone HCl 0.015 mg/kg intravenously single dose (not to exceed 2.4 mg in 4 hours)

☐ morphine sulfate 0.1 mg/kg intravenously single dose (not to exceed 15 mg per dose)

Laboratory UpToDate UpToDate

Blood Bank:

- *Type and crossmatch for potential packed red blood cell (RBC) transfusion are obtained on admission. If at all possible, requested blood should be negative for sickle hemoglobin (Hgb), matched for minor red cell antigens (eg, C, E, Kell), and leukoreduced.*

☐ Type and crossmatch [ ] (blood)

Blood Gases:

- *Arterial blood gas analysis should be considered for patients with significant respiratory distress, in patients that require verification of oxygenation especially if there are inconsistencies between the pulmonary clinical status and recordings from the transcutaneous pulse oximetry, or to provide additional data to determine the need for simple or exchange transfusion (eg, PaO2 <60 mmHg).*

☐ Arterial blood gas (arterial blood)

☐ Venous blood gas (venous blood)

Chemistry:

☐ Basic metabolic panel (serum)

☐ Lactate dehydrogenase (serum)

☐ Comprehensive metabolic panel (serum)

☐ Hepatic function panel (serum)

Hematology:

☐ CBC with platelets and differential (blood)

☐ Reticulocyte count (blood)

Inflammatory Markers:

☐ C-reactive protein (serum)

Microbiology:

- *If the patient is febrile, blood cultures should be obtained.*

☐ Routine culture and sensitivities (blood)

☐ Routine culture and sensitivities (sputum)

Urine:

- *Urinalysis and culture is done in children less than two years with fever and other patients with symptoms suggestive of a urinary tract infection.*

☐ Microscopic examination (urine)

☐ Routine culture (urine)

☐ Urinalysis (urine)

Imaging UpToDate

- *Patients with sickle cell disease (SCD) who have fever, chest pain, or respiratory symptoms should have chest radiographs because acute chest syndrome (ACS) may be present even though it is not clinically suspected (Grade 2C).*

X-Ray:

☐ Routine inspiration PA/lateral X-ray of the chest

Computed Tomography:

☐ Chest CT scan

Consultations

☐ Pediatric Critical Care consultation

☐ Pediatric Hematology consultation

☐ Pediatric Infectious Disease consultation

☐ Pediatric Pulmonology consultation

☐ Pediatrics consultation

Disposition

- *Even the diagnosis of a clinically mild case of acute chest syndrome (ACS) should prompt admission for close monitoring of progressive pulmonary changes, because the clinical status of these patients can quickly deteriorate if the underlying pulmonary insult is not reversed. Managing the child with severe ACS is challenging, and support in an intensive care unit is usually necessary.*

☐ Admit inpatient

☐ Place in observation

☐ Transfer

☐ Discharge patient
