Reason / Problem UpToDate

Sickle Cell Disease Acute Chest Syndrome Pediatric Order Set

- *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. (UpToDate)*
- *The diagnosis of acute chest syndrome (ACS) 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 38.5 degrees C; hypoxemia; and signs of tachypnea, wheezing, cough, or the appearance of increased work of breathing. (UpToDate)*

Sickle Cell Anemia UpToDate UpToDate

Admit / Transfer

- *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. (UpToDate)*

☑ Admit inpatient to Pediatrics as soon as possible

☐ Admit inpatient to Pediatric Intensive Care as soon as possible

☐ Place in observation [Unit] [Observation Duration]

☐ Transfer [Unit]

Condition

☐ Good

☐ Fair

☐ Serious

Code Status:

☐ Full code

Activity

☑ Activity per unit/surgical protocol

☐ Bed rest with bathroom privileges [Duration]

☐ Up ad lib

Diet

☐ Nothing by mouth

☐ Liquid diet, clear liquids only

☐ Special Diet Instructions: encourage oral fluids

☐ Usual diet

Vital Signs

☑ Check vital signs per protocol [Notification Parameter]

☐ Check vital signs [Frequency] [Duration] [Notification Parameter]

IV

- *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.*
- *If the patient is euvolemic and receiving maintenance intravenous fluids, UpToDate suggests using one-quarter or one-half normal saline with or without glucose. Of note, this differs from maintenance fluid replacement in patients without sickle cell disease (SCD), who often receive normal saline. Patients with SCD may have a decreased ability to excrete sodium and may become hypernatremic from receiving normal saline. Hypernatremia in turn may lead to red blood cell dehydration, which increases sickling. (UpToDate)*
- *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. Weights should be monitored daily along with intake/output for assessment of the fluid status and management of the patient. (UpToDate)*

Crystalloid:

☐ Normal saline intravenously [KCl Amount] and [Other Additives] [Rate] [Total Volume]

☐ 5% dextrose in normal saline intravenously [KCl Amount] and [Other Additives] [Rate] [Total Volume]

☐ 5% dextrose in half-normal saline intravenously [KCl Amount] and [Other Additives] [Rate] [Total Volume]

☐ 5% dextrose in quarter-normal saline intravenously [KCl Amount] and [Other Additives] [Rate] [Total Volume]

Crystalloid-Fluid Bolus:

☐ Normal saline intravenous bolus [Bolus Volume] [Rate] [Frequency] [IV Duration]

Blood Products:

- *Numerous case series support the role of transfusion therapy in management of moderate or severe acute chest syndrome (ACS). Transfusion therapy improves oxygenation and should be considered early in the management of these patients. (UpToDate)*

☐ Packed red blood cells, leukocyte reduced (washed / filtered) and crossmatched intravenously [Volume] [Transfusion Duration]

Lock IV:

☐ Saline lock intravenous [Flush Fluid] [Frequency]

Other Nursing

- *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. (UpToDate)*

Assessments:

☑ Assess neurologic signs on admission and then [Frequency] [Notification Parameter]

☑ Complete admission history, pediatric

☑ Complete pediatric pain assessment on admission and then [Frequency]

☑ Obtain weight in kilograms on admission and then [Frequency]

☑ Perform admission assessment pediatric

☐ Measure height in inches on admission and then [Frequency]

Cardiac:

☑ Continuous cardiorespiratory monitoring

Circulatory:

☑ Peripheral IV line care per protocol

☑ Insert peripheral IV line [Qty/Sz/Loc Peripheral IV]

☐ Central venous line care per protocol

Education: UpToDate

- *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)*

☑ Provide disease/medical condition education [Type]

Fluid Balance:

☑ Fluid volume management per protocol

☑ Intake and output on admission and then [Frequency] [Duration]

Respiratory:

☑ Maintain oxygen saturation between greater than or equal to 92% and [Max SpO2 Goal]

☑ Monitor pulse oximetry [Frequency] [Location] [Notify MD if SpO2]

Urinary:

☐ Catheter care per protocol

☐ Insert catheter [Size] [Urinary Catheter] [Frequency]

Therapies

Respiratory Therapy Service: UpToDate

- *Respiratory support, including oxygen supplementation, should be provided to maintain arterial oxygen saturation greater than or equal to 92 percent. Incentive spirometry, preferably supervised by a clinical worker, should be employed at least every two hours to prevent atelectasis from hypoventilation. (UpToDate)*
- *For patients with poor respiratory effort or rising oxygen requirements, the use of non-invasive ventilation, such as nasal mask continuous positive airway pressure (CPAP) or bilevel positive airway pressure (BPAP), may be useful. (UpToDate)*

☐ Respiratory therapy evaluation and treatment

☐ Incentive spirometry [Frequency] only when awake

☐ Warmer , Humidifier oxygen Oxygen Source O2 Rate O2 Sat Goal

Procedures:

☐ Bronchoscopy Proceduralist

☐ Type central venous catheter Proceduralist

☐ Lumbar puncture Proceduralist

Ventilation:

☐ Bilevel positive airway pressure (BPAP)
- Initial IPAP
- Initial EPAP
- Setting Adjustment
- FiO2
- Back Up Respiratory Rate

☐ Continuous positive airway pressure (CPAP)
- Initial CPAP
- Setting Adjustment
- FiO2
- Back Up Respiratory Rate

☐ Non-invasive ventilation
- Mode
- Pressure Support
- Expiratory Pressure
- Respiratory Rate
- Fi02
- Inspiratory Time

Other Therapies: UpToDate

- *A cardinal principle in transfusing individuals with sickle cell disease (SCD) who are critically ill is that exchange transfusion provides greater benefit compared with simple transfusion because only exchange transfusion can significantly lower sickle hemoglobin (HgbS) levels (ie, to less than 30 percent of total Hgb). The lessened effects on viscosity for a given Hgb level are critical in potentially reversing vasoocclusion and improving blood flow. (UpToDate)*

☐ Red blood cell exchange transfusion

Medications

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. (UpToDate)*
- *For patients who are allergic to cephalosporin, clindamycin is given at a dose of 10 to 15 mg/kg IV (maximum single dose 900 mg). (UpToDate)*
- *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). (UpToDate)*

☐ Azithromycin 100 mg/5 mL suspension 10 mg/kg orally single dose (not to exceed 500 mg in 24 hours) *Step 1 - given in addition to ceftriaxone or vancomycin if acute chest syndrome is present.*

☐ Azithromycin 100 mg/5 mL suspension 5 mg/kg orally 1 time per day for 4 days (not to exceed 500 mg in 24 hours) *Step 2 - given 24h after first azithromycin dose.*

☐ cefTRIAXone sodium 75 mg/kg intravenously every 24 hours (not to exceed 2 grams in 24 hours)

☐ Clindamycin phosphate 10 mg/kg intravenously every 6 hours (not to exceed 2.7 grams in 24 hours)

☐ Erythromycin lactobionate 5 mg/kg intravenously every 6 hours (not to exceed 4 grams in 24 hours)

☐ Vancomycin HCl 15 mg/kg intravenously every 6 hours (not to exceed 1 gram in 24 hours)

Antipyretics-Analgesics:

- *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). (UpToDate)*
- *When using ketorolac, one should not simultaneously use other nonsteroidal anti-inflammatory agents (NSAIDs). (UpToDate)*

☐ Ketorolac tromethamine 0.5 mg/kg intravenously single dose as needed for pain for 5 days (not to exceed 30 mg per dose) *Do not combine with other NSAID's.*

☐ Acetaminophen 160 mg/5 mL suspension 15 mg/kg orally every 6 hours as needed for pain or fever (not to exceed 5 doses in 24 hours)

☐ Acetaminophen 650 mg orally every 6 hours as needed for pain or fever (not to exceed 5 doses in 24 hours)

☐ Ibuprofen 100 mg/5 mL suspension 10 mg/kg orally every 6 hours as needed for pain, fever or inflammation (not to exceed 600 mg per dose)

☐ Ibuprofen 400 mg orally every 6 hours as needed for pain, fever or inflammation (not to exceed 600 mg per dose)

Bronchodilators:

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

☐ Albuterol sulfate 90 mcg/actuation aerosol 2 inhalations via metered-dose HFA inhaler every 6 hours as needed for wheezing

☐ Albuterol sulfate 2.5 mg/3 mL nebulizer solution 2.5 mg nebulized every 6 hours as needed for wheezing (not to exceed 5 mg per dose)

☐ Albuterol-ipratropium 2.5-0.5 mg/3 mL nebulizer solution 3 mL nebulized every 6 hours

Corticosteroids:

- *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. (UpToDate)*

☐ methylPREDNISolone 1 mg/kg orally 2 times per day (not to exceed 60 mg in 24 hours)

☐ prednisoLONE 15 mg/5 mL syrup 1 mg/kg orally 2 times per day (not to exceed 60 mg in 24 hours)

☐ predniSONE 1 mg/kg orally 2 times per day (not to exceed 60 mg in 24 hours)

Diuretics:

☐ Furosemide 0.5 mg/kg intravenously single dose as needed for diuresis

Opioid Analgesics: UpToDate

- *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. (UpToDate)*
- *The use of patient controlled analgesia (PCA) may minimize oversedation and hypoventilation but still provide adequate pain control. (UpToDate)*

☐ oxyCODONE 5 mg/5 mL solution 0.1 mg/kg orally every 6 hours as needed for moderate to severe pain (not to exceed 10 mg per dose)

☐ oxyCODONE HCl 5 mg orally every 6 hours as needed for moderate to severe pain

☐ HYDROmorphone HCl 0.015 mg/kg intravenously every 4 hours as needed for moderate to severe pain (not to exceed 1.2 mg in 4 hours)

☐ HYDROmorphone HCl (0.2 mg/mL concentration) via intravenous patient controlled analgesia

- Loading Dose
- Basal Rate
- Demand Dose
- Lockout Interval
- Max Dose Per 1 Hour
- Max Dose Per 4 Hours

☐ morphine sulfate 0.1 mg/kg intravenously every 2 hours as needed for moderate to severe pain (not to exceed 4 mg per dose)

☐ morphine sulfate via intravenous patient controlled analgesia

- Loading Dose
- Basal Rate
- Demand Dose
- Lockout Interval
- Max Dose Per 1 Hour
- Max Dose Per 4 Hours

Prophylactic Agents: UpToDate

- *For patients with sickle cell disease (SCD) over the age of 18 to 24 months with one or more of the following problems, UpToDate recommends hydroxyurea in addition to symptomatic management and transfusion, rather than symptomatic management and transfusion alone: frequent painful episodes; history of acute chest syndrome; history of other severe vasoocclusive events; severe symptomatic anemia. (UpToDate)*
- *Hydroxyurea should be administered by experienced clinicians with close medical supervision and monitoring for potential toxicity. (UpToDate)*

☐ Folic acid 1 mg orally 1 time per day

☐ Hydroxyurea 15 mg/kg orally 1 time per day

☐ Multivitamins 1 tablet orally 1 time per day

☐ Penicillin V potassium 125 mg/5 mL suspension 125 mg orally 2 times per day *Patient age 3 months to 2 years.*

☐ Penicillin VK 250 mg/5 mL suspension 250 mg orally 2 times per day *Patient age 2 to 5 years.*

Vaccines: UpToDate

☐ Influenza virus vaccine trivalent split (equivalent to Fluzone) 0.25 mL intramuscularly single dose *Age 6 to 35 months.*

☐ Influenza virus vaccine trivalent split (equivalent to Fluzone) 0.5 mL intramuscularly single dose *Patients greater than 3 years of age.*

☐ Pneumococcal 13-valent conjugate vaccine 0.5 mL intramuscularly single dose

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Laboratory 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. (UpToDate)*

☑ Type and crossmatch (blood) # of Units

☐ Type and screen (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 less than 60 mmHg). (UpToDate)*

☑ 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)

☐ Renal function panel (serum)

Hematology:

- *Laboratory tests should be obtained promptly and include blood culture, complete blood count, and reticulocyte count. (UpToDate)*

☑ CBC with platelets and differential (blood)

☑ Reticulocyte count (blood)

Microbiology:

- *If the patient is febrile, blood cultures should be obtained. Culture of other sites (eg, urine, stool, sputum) should be obtained as clinically indicated. (UpToDate)*

☐ Routine culture and sensitivities # of Sets (blood)

☐ Routine culture and sensitivities # of Sets (sputum)

CSF:

- *Cerebrospinal fluid is obtained for analysis and culture only if there is a clinical suspicion for meningitis. (UpToDate)*

☐ Cell count with differential (cerebrospinal fluid)

☐ Glucose (cerebrospinal fluid)

☐ Gram stain (cerebrospinal fluid)

☐ Protein (cerebrospinal fluid)

☐ Routine culture and sensitivities # of Sets (cerebrospinal fluid)

Inflammatory Markers:

☐ C-reactive protein (serum)

Urinalysis:

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

☐ Urinalysis (urine)

☐ Microscopic examination (urine)

☐ Routine culture and sensitivities # of Sets (urine)

Imaging

- *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). (UpToDate)*
- *In patients with acute chest syndrome (ACS) and progressive respiratory difficulty where the cause remains obscure, the possibility of pulmonary embolus should be considered especially in patients who are rapidly deteriorating with worsening hypoxia, increasing A-a gradient, and/or signs of cardiogenic dysfunction. (UpToDate)*

X-Ray:

☐ Routine inspiration PA/lateral X-ray of the chest today [Reason for X-ray]

Computed Tomography:

☐ Chest CT scan with IV contrast today [Reason for Study]

Other Tests

Pulmonary Testing:

☐ Pulmonary function testing today

Consultations

☐ Pediatric Critical Care consultation today [Reason for Consultation]

☐ Pediatric Hematology consultation today [Reason for Consultation]

☐ Pediatric Infectious Disease consultation today [Reason for Consultation]

☐ Pediatric Pulmonology consultation today [Reason for Consultation]

☐ Pediatrics consultation today [Reason for Consultation]
