ACUTE CHEST SYNDROME – GUIDELINES FOR MANAGEMENT

Background: Acute chest syndrome is an important cause of morbidity and mortality in patients with sickle cell disease. More than 50% of patients will have at least one episode during childhood. Patients experiencing repeated episodes are at high risk for chronic lung disease and early death.

Definition: Acute chest syndrome in a patient with sickle cell disease is defined as a new infiltrate on CXR, accompanied by at least one of the following – chest pain, fever, hypoxia or respiratory distress (such as shortness of breath, wheezing or persistent cough). Unfortunately, the other findings may appear prior to CXR changes. Therefore, a normal CXR initially will not always rule out this condition.

Management:
1. Chest X-rays: If the patient’s first CXR shows no new infiltrates, do not assume that acute chest syndrome has been ruled out. If the other typical symptoms (eg.chest pain) continue, or if the patient’s clinical status worsens, repeat the CXR within 24 hours. If a patient has chest pain as his only symptom, and if he is sent home on oral pain medication, a follow-up should be made for the next day – at a minimum a phone assessment must be made the next day.

2. Antibiotics: If the patient has a fever > 101°F, start antibiotic treatment. Use a combination of IV ceftriaxone and oral azithromycin. This combination should cover Pneumococcus, Chlamydia and Mycoplasma.

3. Fluid Management: If the patient already meets the criteria for acute chest syndrome, or if it is being seriously considered - take care to avoid over hydration. Patients should receive only maintenance IV fluids plus what they want to drink, unless they are clinically dehydrated. They should not receive large IV boluses. There is evidence that pulmonary edema can lead to rapid respiratory decompensation in patients with acute chest syndrome.

4. Pain Management: If the patient complains of chest pain, give NSAIDS on a regular schedule (Ibuprofen 10 mg/kg PO Q6 hours or Toradol 0.5 mg/kg IV Q6 hours). Many patients will need narcotics in addition. Follow the pain management protocol. Good pain control will help if the patients are not taking deep breaths due to splinting from pain. At the same time, these patients need to be monitored closely for respiratory depression, as it can be counter-productive too. Consider titrating pain management to an effective regimen – that is managing pain and at the same time is not sedating.

5. Transfusion: A patient with acute chest syndrome (documented infiltrates or normal CXR with high clinical suspicion) with respiratory distress (tachypnea, retractions or hypoxia), should be transfused with PRBCs, aiming for a Hgb level of ~ 10gms/dL. It may take more than one transfusion of 10cc/kg to achieve that level.

Exchange transfusion is considered for patients with rapidly worsening respiratory status who have already received simple transfusions to bring their Hgb ~ 10gms/dL or whose hemoglobins are higher > 9.5 gms/dl to begin with, that they cannot be given a simple transfusion due to risk of increasing hyperviscosity. When transfusing PRBCs – order should be for sickledex negative, leukocyte depleted and cross-matched for C, E and Kell antigens.

6. Oxygen: Monitor by continuous pulse oximetry. Administer O₂ to maintain saturations >92%. Consider delivering oxygen by face mask rather than by nasal prongs in patients who are obligate mouth breathers because of tonsillar hypertrophy. If respiratory distress is worsening – use of positive pressure ventilation - BiPAP should be also considered in addition to the blood transfusions and antibiotics.

7. Incentive Spirometry: Encourage patients to participate in incentive spirometry Q 2 hours during day and Q 4hours overnight to maximize alveolar expansion. Post a schedule so that this is not missed.

8. Prevention of Gastritis: Several of the drugs recommended above may cause gastritis. Therefore it is appropriate to start H₂ blockers routinely in these patients.

9. Bronchodilators: Consider trial of bronchodilators, especially in patients with history of reactive airway disease or wheezing on exam. For patients who have asthma/reactive airway disease continue home asthma action plan.

10. Clinical deterioration: It is important to remember that acute chest syndrome can progress very quickly if not promptly recognized or treated. In severe cases, it can also progress despite measures including blood transfusions, antibiotics etc. Patients with a diagnoses or clinical suspicion of acute chest syndrome should be monitored closely. If respiratory distress/hypoxia continues to get worse despite straight blood transfusion and antibiotics – consider transfer to PICU for close monitoring, increased respiratory support and possible exchange transfusion. Inform PICU staff if respiratory status appears to be worsening despite the strategies outlined above. Make patient NPO – as an exchange transfusion will require a pheresis catheter placement and sedation. Goal of exchange transfusion will be to bring Hgb S< 30%.
Respiratory support with BiPAP/mechanical ventilation may be required. We can consider dexamethasone for refractory acute chest.

11. Lab Monitoring: Patients will need daily CBC’s and reticulocyte counts. Many patients experience a precipitous fall in Hgb during an episode of acute chest syndrome. If symptoms worsening – consider CMP to monitor for electrolyte abnormalities, kidney and liver function tests.

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