2. ACUTE CHEST SYNDROME

Principles

- Acute chest syndrome (ACS) is a potentially life-threatening complication of sickle cell disease requiring a high index of suspicion.
- ACS-related complications may be reduced by prompt intervention and appropriate therapy , including the use of blood transfusion.

Recommendations

Early Detection and Prevention

- Pulse oximetry should be a routine part of every medical assessment to identify changes from a patient's baseline oxygen saturation level.[9]
- Patients admitted for a *febrile* or *vaso-occlusive episode* should be carefully examined and have continuous oxygen saturation monitoring for early detection of clinical symptoms and/or changes in oxygenation.
- Hydroxyurea therapy should be considered for patients after a single episode of acute chest syndrome.[10] (*see Part I, section 1 on Hydroxyurea*)
- There is conflicting evidence about the benefit of chronic red blood cell (RBC) transfusions for the prevention of ACS, and they and are not routinely recommended.[11,12]
- Asthma management for children and adults with both sickle cell disease and asthma should be optimized, and a referral to an asthma specialist should be considered.

Diagnosis and Management

- Once ACS diagnosis is established:
  - Admit to hospital for close care and monitoring.
  - Administer high-flow oxygen to maintain an oxygen saturation of ≥95%.
  - Identify and treat airway, breathing, and circulation issues.
  - Maintain hydration with intravenous and oral intake to keep the total fluid intake at maintenance rate. Monitor fluid balance closely, as overhydration is associated with pulmonary edema and worsened clinical status.[13]
  - Treat pain aggressively by giving adequate analgesia while monitoring oxygen saturation and blood carbon dioxide, as opiates can cause hypoventilation and worsening oxygen saturation. (*see Part II, section 1 on Pain*).
  - Obtain a complete blood cell count, reticulocyte count, arterial blood gases, and extended cross match for possible exchange or simple transfusion.
  - For febrile patients, obtain blood cultures and consider broad-spectrum antibiotics (including a macrolide for children at risk of atypical bacterial infections).
  - Acute pulmonary embolism should also be considered in the differential diagnosis for at-risk patients, as the clinical presentations can be similar.
  - Consult Hematology service urgently prior to transfusion or admission.
  - The use of corticosteroids for the management of ACS has yielded equivocal results, and is not routinely recommended.
  - Patients should use incentive spirometry and/or be assessed by a respiratory therapist.
  - Oxygen saturation and oxygen requirements should be monitored closely.
  - Patients should be reviewed early by a critical care outreach team, if available, if there are concerns about serious or worsening clinical status.

Background

ACS is an acute complication of sickle cell disease defined as the presence of a new pulmonary infiltrate (involving at least one complete lung segment and not atelectasis), fever, chest pain, and/or respiratory signs and symptoms. Clinical signs and symptoms overlap with those of a lower respiratory tract infection. ACS is a leading cause of hospitalization in patients with sickle cell disease, and a significant risk factor for early mortality.

The incidence of ACS is highest among patients with HbSS genotype, and is more common in children than in adults. Causes of ACS include infections (bacteria, viruses, Mycoplasma, Chlamydia and mixed infections), pulmonary infarction, and fat embolism. Acute pulmonary embolism should also be considered in the differential diagnosis in a patient presenting with apparent ACS, as the clinical presentations can be similar. Patients with sickle cell disease who have undergone abdominal surgery are at risk of developing acute chest syndrome in the immediate post-operative period. Children with sickle cell disease and comorbid asthma are twice as likely to develop ACS as those with sickle cell disease who do not have asthma.[3] Age, hemoglobin F level, degree of anemia, and a higher steady-state white blood cell (WBC) count are also independent risk factors for ACS.3 In patients with hemoglobin SC disease (HbSC), only WBC count was found to be a statistically significant risk factor.[1]

Up to 50% of hospitalized patients with ACS are admitted for other reasons – most commonly vaso-occlusive crises – and develop ACS during admission.[1] Clinical signs and symptoms of acute chest syndrome are similarin both HbSS and HbSC genotypes.[2] At the time of diagnosis, the most frequent symptoms of ACS include fever, cough, chest pain, shortness of breath, and tachypnea.[4] Other symptoms include wheezing, chills, abdominal pain, rib pain, and pain in the arms and legs. Fever and cough are more common in young children, while chest pain, productive cough, and hemoptysis are more common in adult patients. Neurologic complications may also occur. Symptoms observed during a patient's first episode of acute chest syndrome are generally predictive of symptoms during subsequent events.

It is important to note that recurrent ACS is associated with an increased risk of stroke.[1,5] Recurrent ACS is also associated with an increased risk of pulmonary hypertension, interstitial lung disease and pulmonary fibrosis.[6-8]

References

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