Without a Transfusion – SANGUINATE and the Management of Acute Chest Syndrome

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Learning Objectives
- Describe the pathophysiology of acute chest syndrome
- Recognize the important role of blood transfusion in acute chest syndrome
- Identify additional therapies for those not able to receive blood transfusion, specifically SANGUINATE.

Patient Presentation
29 year old African-American female with Sickle Cell Disease
- 1 day history of left sided chest pain, cough productive of yellow-green sputum, and a temperature of 38.3°C
- Jehovah’s Witness – Refusing Blood Transfusions

Physical Exam:
HR: 120 BP: 121/70 RR: 28 SpO2: 92% on 3L
Skin: Pale conjunctiva with dry mucous membranes
Respiratory: Left lower lobe crackles, no wheezes or rhonchi
Cardiac: Tachycardic, III/VI SEM in left upper sternal border

Laboratory Data:
Chest X-ray showed left lower lobe infiltrate concerning for an infectious process
Sputum cultures and blood cultures showed no growth

Diagnosis of Acute Chest Syndrome
- Diagnosis is made on average 2.5 days after initial presentation
- New segmental radiographic evidence of a pulmonary infiltrate at least one of the following:
  - Temperature ≥38.5ᴼC
  - >2% decrease in O₂ saturation from a documented steady-state value on room air
  - PaO₂ <60 mmHg
  - Tachypnea (per age of 3 years old)
  - Intercoastal retractions, nasal flaring, or use of accessory muscles of respiration
  - Chest pain
  - Cough
  - Wheezing or Rales

Hospital Course

Day 1
- Admitted
- Increasing tachypnea and tachycardia

Day 2
- Transferred to ICU
- Oxygen requirement improves, tachycardia resolves

Day 3
- Receives 4 units of SANGUINATE
- Tachycardia and dyspnea improves

Day 4 - 9

Day 10
- Oxygen requirement improves, tachycardia resolves

Day 15
- Day 23
- Discharged home

Treatment of Acute Chest Syndrome
- Reduce transfusion is the standard of care
  - Empiric Antibiotic therapy, usually with Azithromycin
  - Supplemental Oxygen
  - Pain Control
  - IV Fluid Hydration

References

Take Home Points
1. Acute Chest Syndrome is a life-threatening pulmonary condition in Sickle Cell patients
2. The standard of care for treatment of Acute Chest Syndrome is exchange transfusion
3. SANGUINATE is a promising alternative to blood transfusion in patients who are unable or unwilling to receive blood products