ACUTE CHEST SYNDROME

TERM DEFINITION

Acute chest syndrome (ACS) is defined as an acute illness in a patient with sickle cell disease characterized by: 1) fever and/or respiratory symptoms, and 2) new pulmonary infiltrate on chest X-ray.

RISK FACTORS

In about 50% of cases, acute chest syndrome (ACS) develops one-three days **following onset of a vaso-occlusive crisis**.

ACS can also develop **after surgery**, especially following abdominal surgery and in cases where patients do not receive appropriate pre-operative blood transfusion.

CLINICAL PEARLS

Second most frequent reason for hospitalization in children and adults with sickle cell disease (SCD).

Most common cause of death in children and adults with SCD.

Variable clicnial spectrum from a mild pneumonic illness to acute respiratory distress syndrome and multi-organ failure.

HbSS > HbSC

Its incidence in HbSC disease and HbS-β+ thalassemia is less than in sickle cell anemia (HbSS and HbS-β0 thalassemia).

ACS occurs and can be severe in all sickle genotypes.

Most common bacterial organism identified in adults is *chlamydophila pneumoniae*.

Hemoglobin concentration often reduced vs. baseline.

Hemolysis markers often increased vs. baseline.

Radiological signs often lag behind the physical signs.

DIAGNOSIS

Diagnosis of acute chest syndrome (ACS) is based on the finding of recent onset of fever, tachypnea, chest signs, and hypoxia in a patient with sickle cell disease (SCD) and the appearance of consolidation on a plain chest X-ray.

CLINICAL PRESENTATION

- **Cough** - may be productive.
- **Chest pain** - may be pleuritic.
- Shortness of breath
- Rib and sternal pain
- Chills
- Wheezing
- Hemoptysis
- Fever
- Tachypnea
- Tachycardia
- Wheezing
- Additional chest signs
- Low oxygen saturation

LABS

- **Complete blood count** - may show decreased Hb and platelets from baseline.
- Renal and liver function
- **Hemolysis markers** - may be increased from baseline.
- Blood group and screen.
- Arterial blood gases if SpO2 < 95% on room air
- Blood cultures
- Serology for atypical respiratory organisms
- Urine for Pneumococcal and Legionella antigen
- Sputum for bacterial culture
- **CXR** - typical findings are segmental, lobar or multilobar consolidation usually involving the lower lobes +/- pleural effusion.

TREATMENT

SUPPLEMENTAL OXYGEN

- Maintain SpO2 ≥ 95% or within 3% of the patient's baseline.
- Monitor SpO2 at least 4-hourly to detect clinical deterioration indicated by increasing oxygen requirement.

IV FLUIDS

- Intravenous crystalloid infusion should be administered until the patient can drink adequate amounts of fluid.
- Fluid requirements should be individualized and be guided by the patient's fluid balance and cardiopulmonary status.

TRANSFUSION

- Simple transfusion if PaO2 of less than 90 kPa on room air, aim for Hb 10-11 g/dL.
- Exchange transfusion in patients with severe disease* or those with Hb.>.9 g/dL. Aim for Hb 10-11 g/dL and HbS < 30%-40%.

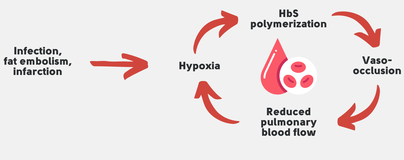
OTHER

- Pain management
- Incentive spirometry
- Antibiotics - since it is not possible to reliably rule out an infectious etiology of the ACS, all patients should be treated with empiric antibiotics for severe community acquired pneumonia.

*SpO2 < 90% despite supplemental O2, increased respiratory distress, progressive pulmonary infiltrates, and/or not responding to simple transfusion

PROXIMATE MECHANISMS

The initial insult, which may include pulmonary infection, fat embolism and/or pulmonary infarction, causes a **reduction in alveolar oxygenation tension**, which, in turn, leads to **HbS polymerization**. Sickling results in **decreased pulmonary blood flow** that exacerbates vaso-occlusion, producing more severe hypoxia such that a **vicious cycle** of hypoxia, HbS polymerization, vaso-occlusion and altered pulmonary blood flow ensues. In many cases, the specific cause or inciting factor is not apparent.



HISTORY OF MEDICINE

History of SCD dates back to James Herrick's original description of the disease in 1910, followed by Linus Pauling's discovery of a modified form of hemoglobin in blood of SCD patients, leading Pauling and others to label SCD as the first molecular disease, and Vernon Ingram's identification of the amino acid (aa) substitution in the beta chain of sickle Hb.

7,000 years ago
Estimated origin of mutation

1910
James Herrick
1st description of SCD

1949
Linus Pauling
SCD: Modified form of Hb (1st "molecular disease")

1956
Vernon Ingram
AA substitution

1998
FDA approval Hydroxyurea

NOTES

ATTRIBUTIONS

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The Blood Project
ENCYCLOPEDIA OF BLOOD