

Sickle Cell Disease and Its Complications: Focus on Acute Chest Syndrome

Miguel Nascimento*

Department of Health and Life Science, University of Rio de Janeiro, Rio de Janeiro, RJ, Brazil

INTRODUCTION

Sickle Cell Disease (SCD) is a hereditary blood disorder characterized by the production of abnormal hemoglobin, leading to distorted red blood cells that can cause various complications. Among these complications, Acute Chest Syndrome (ACS) stands out as a severe and potentially life-threatening condition

Sickle cell disease affects millions worldwide, primarily individuals of African, Mediterranean, Middle Eastern and Indian ancestry. The disorder arises from a mutation in the β -globin gene, resulting in the production of Hemoglobin S (HbS). Under low oxygen conditions, HbS polymerizes, leading to the characteristic sickle shape of red blood cells. This shape can cause vaso-occlusion, resulting in ischemia, pain crises and organ damage.

Acute chest syndrome, a significant complication of SCD, is characterized by the sudden onset of chest pain, fever and respiratory symptoms such as cough, difficulty breathing and hypoxia. It is often precipitated by infection, pulmonary embolism or vaso-occlusive events.

DESCRIPTION

Risk factors for acute chest syndrome

Infection: Infections are a leading cause of ACS. Patients with SCD are more susceptible to respiratory infections due to functional asplenia, often resulting from repeated splenic infarctions. Common pathogens include *Streptococcus pneumoniae*, *Haemophilus influenzae* and increasingly, viral infections like influenza and COVID-19. Vaccinations and prophylactic antibiotics are crucial for prevention.

Pulmonary complications: SCD can lead to chronic pulmonary complications, including asthma and pulmonary hypertension, which increase the risk of ACS. Acute lung injury may also arise from inflammatory processes triggered by vaso-occlusion or infection.

Hypoxia and dehydration: Situations that lower oxygen availability, such as high altitudes, strenuous exercise or dehydration, can provoke sickling and increase the likelihood of ACS. Education on avoiding these triggers is vital for patients.

Vaso-occlusive crises: Painful crises resulting from blood vessel blockage can lead to ACS. Vaso-occlusion in pulmonary circulation can induce local ischemia, triggering inflammation and subsequent chest syndrome.

Chronic conditions: Comorbidities like obesity and diabetes can exacerbate the risks associated with ACS by contributing to overall health decline and increasing susceptibility to infections.

Clinical presentation of acute chest syndrome

The clinical presentation of ACS can vary, but it typically includes:

Chest pain: Often localized but can be diffuse, presenting as pleuritic or non-pleuritic pain.

Respiratory symptoms: Patients may experience cough, dyspnea and wheezing.

Fever: Often indicative of an underlying infection, with temperatures usually above 38°C (100.4°F).

Hypoxemia: Low oxygen saturation levels are commonly observed, necessitating prompt intervention.

A chest X-ray is essential for diagnosing ACS, revealing new pulmonary infiltrates that may indicate infection, infarction or edema.

Management of acute chest syndrome

Management of ACS involves a multidisciplinary approach, including the following components:

Hospitalization: Patients presenting with ACS typically require hospitalization for close monitoring and treatment.

Oxygen therapy: Supplemental oxygen is crucial to address hypoxemia. Maintaining oxygen saturation above 92% is a primary goal.

Correspondence to: Miguel Nascimento, Department of Health and Life Science, University of Rio de Janeiro, Rio de Janeiro, RJ, Brazil; E-mail: minguelnm@edu.br

Received: 28-Oct-2024, Manuscript No. JGSGT-24-34854; **Editor assigned:** 02-Nov-2024, PreQC No. JGSGT-24-34854 (PQ); **Reviewed:** 16-Nov-2024, QC No. JGSGT-24-34854; **Revised:** 13-Jun-2025, Manuscript No. JGSGT-24-34854 (R); **Published:** 20-Jun-2025, DOI: 10.35248/2157-7412.25.16.460

Citation: Nascimento M (2025) Sickle Cell Disease and Its Complications: Focus on Acute Chest Syndrome. J Genet Syndr Gene Ther. 16:460.

Copyright: © 2025 Nascimento M. This is an open-access article distributed under the terms of the Creative Commons Attribution License, which permits unrestricted use, distribution and reproduction in any medium, provided the original author and source are credited.

Hydration: Intravenous fluids are often necessary to promote hydration, prevent vaso-occlusion and support overall circulatory function.

Pain management: Effective analgesia is essential to manage pain and improve patient comfort. Opioids are commonly used, but careful monitoring is required due to the risk of respiratory depression.

Antibiotics: Empirical broad-spectrum antibiotics should be initiated promptly to address potential infections. Coverage typically includes respiratory pathogens, with adjustments made based on culture results.

Transfusion therapy: In cases of severe ACS or significant anemia, blood transfusions may be necessary to increase the proportion of normal red blood cells, thereby reducing sickling and improving oxygen delivery.

Incentive spirometry: Patients should be encouraged to use incentive spirometry to promote deep breathing, prevent atelectasis and improve lung function.

Supportive care: Supportive measures, including physical therapy and respiratory support, can enhance recovery and prevent complications.

Long-term considerations and prevention strategies

To reduce the risk of ACS, long-term management of SCD patients should focus on prevention and education. Key strategies include:

Routine screenings: Regular health check-ups to monitor for complications, including pulmonary hypertension and asthma, can facilitate early intervention.

Vaccinations: Vaccines against pneumonia, influenza and meningitis are critical for preventing infections that could lead to ACS.

Patient education: Educating patients about recognizing early symptoms of ACS and managing triggers is essential for improving outcomes. Patients should be aware of the importance of hydration, avoiding high altitudes and recognizing the signs of infection.

Comprehensive care: A multidisciplinary approach involving hematologists, pulmonologists and primary care providers can ensure comprehensive management of SCD and its complications.

Research and new therapies: Ongoing research into new therapies, including gene therapy and novel medications, holds promise for reducing the burden of SCD and its complications, including ACS.

CONCLUSION

Acute chest syndrome is a significant and potentially life-threatening complication of sickle cell disease. Understanding its risk factors, clinical presentation and management strategies is crucial for healthcare providers and patients alike. By prioritizing prevention, early recognition and effective management, we can improve outcomes for individuals living with SCD and reduce the incidence of ACS. Comprehensive care that includes education, routine screenings and multidisciplinary collaboration will be key in enhancing the quality of life for affected patients. It is critical to understand the risks associated with ACS in patients with SCD to enhance prevention, early detection and management strategies.