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Acute Chest Syndrome in Patients with Sickle Cell Disease in Nigeria: Clinical Features and Management

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Abstract

Acute chest syndrome (ACS) is one of the most serious pulmonary complications of sickle cell disease and remains an important cause of hospitalization and death among affected patients. This issue is particularly relevant in Nigeria, where sickle cell disease is common and access to early diagnosis, blood transfusion, respiratory support, and specialist care may differ between medical settings. ACS does not usually result from a single cause. Pulmonary vaso-occlusion, infection, fat embolism, hypoventilation, and tissue hypoxia may occur together and lead to rapid deterioration. Patients may initially present with fever, cough, chest pain, tachypnea, or mild hypoxemia, while severe cases can progress to multilobar pulmonary involvement, marked oxygen desaturation, respiratory failure, and other systemic complications. The clinical picture may also differ between children and adults, which makes early recognition especially important. Diagnosis is mainly based on respiratory symptoms combined with a new pulmonary infiltrate on chest imaging, supported by laboratory findings and assessment of oxygenation. Clinical management requires more than treatment of the pulmonary lesion alone. Oxygen therapy, antibiotics, appropriate pain control, cautious fluid therapy, simple blood transfusion, exchange transfusion, and respiratory support may all be needed depending on disease severity. In Nigeria, delayed presentation, limited access to blood products, uneven availability of exchange transfusion, and restricted diagnostic resources may further complicate care. Better recognition of high-risk patients, earlier treatment, standardized hospital management, and improved long-term sickle cell disease care are therefore important for reducing severe ACS-related outcomes.

Keywords: sickle cell disease, acute chest syndrome, pulmonary complications, clinical management

1. Introduction

Sickle cell disease (SCD) is an inherited hemoglobin disorder characterized by the presence of abnormal hemoglobin S (HbS). Under conditions such as hypoxia, dehydration, or infection, HbS can polymerize and make red blood cells more rigid. These altered cells may obstruct small blood vessels and contribute to vaso-occlusion, hemolysis, tissue ischemia, and repeated organ injury. Acute chest syndrome (ACS) is one of the most serious pulmonary complications of SCD and can develop during both childhood and adulthood.

ACS is generally defined by a new pulmonary

infiltrate on chest imaging together with clinical features such as fever, cough, chest pain, tachypnea, dyspnea, or hypoxemia. Its presentation is not always obvious at the beginning. Some patients are admitted for pain or fever and develop respiratory signs only later. Infection, pulmonary vaso-occlusion, fat embolism, and hypoventilation may all contribute to its development, which also explains why the clinical course can differ considerably between patients (Adegoke et al., 2024).

The condition is especially relevant in Nigeria because SCD is common and respiratory complications remain an important cause of hospital admission. In a multicenter retrospective study involving ten tertiary hospitals in Nigeria, Ibraheem et al. (2024) reviewed 7,256 hospital admissions among children and adolescents with SCD. Respiratory morbidities were identified in 1,213 cases, and ACS accounted for 26.7% of lower respiratory presentations. Of the 17 respiratory-related deaths recorded in the study, 11 occurred in patients with ACS. These findings show that ACS is not only a frequent complication in Nigerian clinical practice but also an important contributor to severe outcomes.

The Nigerian setting also affects how ACS is recognized and managed. Patients may present to hospitals with different levels of access to imaging, oxygen therapy, blood transfusion, and specialist hematology care. For this reason, early recognition is particularly important. The Paediatric Association of Nigeria has developed specific guidance for ACS in children with SCD, adapting some recommendations to local clinical conditions (Adegoke et al., 2024). This local approach provides an important basis for discussing the clinical features, diagnosis, and management of ACS in Nigerian patients.

2. Pathogenesis and Major Triggers of Acute Chest Syndrome

Acute chest syndrome develops through several interacting mechanisms rather than a single pathway. Pulmonary vaso-occlusion is one of the central processes. When oxygen tension falls, hemoglobin S polymerizes more easily, sickled red blood cells become less flexible, and blood flow through the pulmonary microcirculation is impaired. This can lead to local ischemia, endothelial injury, inflammation, and further hypoxemia, creating a cycle that may rapidly worsen respiratory function.

Infection is another major trigger, especially in children. Respiratory infection can produce fever, inflammation, and local hypoxia, all of which increase the tendency of red cells to sickle. Vichinsky et al. (2000) identified a range of bacterial, viral, and atypical pathogens in patients with ACS, although a definite infectious cause was not found in every episode. In Nigeria, infection is also clinically important because it can precipitate vaso-occlusive events through fever, dehydration, inflammatory responses, and changes in oxygenation (Ahmed, 2011).

Fat embolism is more often associated with severe vaso-occlusive episodes and is particularly relevant in older adolescents and adults. Bone marrow ischemia and necrosis can release fat droplets into the circulation, which may then reach the lungs and contribute to pulmonary injury. ACS can also develop after severe pain because chest or abdominal pain may limit deep breathing. Opioid-related sedation, surgery, or prolonged immobility may further reduce ventilation and promote atelectasis.

Other factors such as asthma, dehydration, and recent surgery can increase risk, but they usually act together with the main mechanisms rather than independently. In Nigerian children with SCD, a history of asthma has been associated with ACS, suggesting that pre-existing airway disease may make pulmonary complications more likely (Adegoke et al., 2015). Overall, ACS is best understood as the result of overlapping vaso-occlusion, infection, embolic injury, and impaired ventilation, with the importance of each factor varying between patients.

3. Clinical Features and Disease Progression

The clinical presentation of acute chest syndrome varies between patients. Common features include fever, cough, chest pain, tachypnea, dyspnea, hypoxemia, and sometimes wheezing. These findings are not specific to ACS, and early disease may resemble pneumonia or an uncomplicated vaso-occlusive crisis. Some patients develop clear respiratory symptoms at admission, while others are hospitalized mainly for pain or fever and show pulmonary involvement later.

Age influences the clinical picture. In a large study of patients with sickle cell disease, younger children more often presented with fever and cough, whereas adults were more likely to have shortness of breath, chills, severe pain, multilobar pulmonary involvement, and pleural effusion

(Vichinsky et al., 1997). Adults also tended to have longer hospital stays and more severe outcomes. This difference is clinically relevant because ACS in children may initially look like an acute respiratory infection, while in adults it may emerge during a severe painful crisis.

Chest examination can reveal crackles, wheezing, reduced breath sounds, or signs of consolidation, but these findings may be limited early in the illness. Oxygen saturation is therefore an important part of assessment, especially when it falls below the patient's usual level. Laboratory findings are also variable. Hemoglobin may decrease during an episode, while leukocytosis is common but nonspecific. Imaging usually shows a new pulmonary infiltrate, which may remain localized or extend to several lobes as the disease progresses (Vichinsky et al., 1997).

The course of ACS can change quickly. A patient who initially has mild respiratory symptoms may later develop increasing oxygen requirements, persistent tachypnea, more extensive infiltrates, and respiratory distress. Severe disease may progress to respiratory failure, neurological abnormalities, hemodynamic instability, or dysfunction of other organs. The direction of change is often more important than any single finding at admission.

A Nigerian case reported by Alli et al. (2012) illustrates this pattern. A 28-year-old man with sickle cell anemia was admitted with fever, severe chest pain, and a hemolytic crisis. He initially received antibiotics, intravenous fluids, analgesia, and blood transfusion. About 72 hours later, however, he developed severe hypoxemia and required transfer to intensive care, where oxygen therapy and circulatory support were provided. He later recovered. The case shows that ACS may develop or worsen after hospitalization rather than presenting as a complete respiratory syndrome from the beginning.

Broader Nigerian data support the same concern. In a multicenter study involving ten tertiary hospitals, Ibraheem et al. (2024) found that ACS was an important respiratory complication among children and adolescents with SCD and accounted for most of the respiratory-related deaths recorded in the study. Respiratory mortality was also more closely associated with the early period of hospitalization. These findings reinforce the need to pay close attention to new respiratory symptoms during the first days of admission.

Clinical progression should be suspected when oxygen saturation falls, respiratory rate continues to increase, pulmonary infiltrates become more extensive, or the patient develops greater work of breathing and fatigue. Previous ACS, frequent vaso-occlusive crises, asthma, and extensive pulmonary involvement may increase concern, but no single feature can predict the course in every patient. ACS is therefore best viewed as a dynamic complication whose severity may change over hours rather than a fixed diagnosis established at admission.

4. Diagnosis and Clinical Assessment

The diagnosis of acute chest syndrome is based on clinical symptoms together with a new pulmonary infiltrate on chest imaging. In patients with sickle cell disease, ACS should be suspected when fever, cough, chest pain, tachypnea, dyspnea, or hypoxemia appears, especially when these symptoms develop during hospitalization for a vaso-occlusive crisis. Because early manifestations may be mild or nonspecific, repeated assessment is often more useful than relying on a single examination (Howard et al., 2015; Adegoke et al., 2024).

Chest radiography remains the main imaging method for diagnosis. Pulmonary infiltrates may be focal or multilobar and can become more extensive as the disease progresses. Pulse oximetry is also important because a decline in oxygen saturation may indicate worsening pulmonary involvement. Basic laboratory tests usually include hemoglobin, white blood cell count, platelet count, and other routine indicators. These findings are most useful when compared with the patient's baseline values rather than interpreted separately. In patients with marked hypoxemia or respiratory distress, arterial blood gas analysis and further imaging may be considered.

Differential diagnosis can be difficult because ACS overlaps with several other conditions. Pneumonia is particularly important because infection may both resemble and trigger ACS. Pulmonary embolism, asthma exacerbation, atelectasis, pulmonary edema, and fat embolism should also be considered according to the clinical picture. Identifying one of these conditions does not always exclude ACS, since more than one process may be present at the same time (Vichinsky et al., 2000).

Clinical assessment should also determine

disease severity. Persistent hypoxemia, increasing oxygen requirement, multilobar infiltrates, marked tachypnea, falling hemoglobin, altered mental status, or hemodynamic instability suggest a more serious course. These findings should prompt closer monitoring and early escalation of care. In practice, the most important issue is not only whether the patient meets the diagnostic definition of ACS, but whether respiratory function is worsening over time.

5. Clinical Management of Acute Chest Syndrome

The management of acute chest syndrome should begin as early as possible because respiratory deterioration may occur even when the initial presentation appears moderate. Treatment usually combines oxygen therapy, antimicrobial treatment, pain control, careful fluid management, transfusion, and respiratory support according to disease severity. The main aim is to maintain adequate oxygenation while reducing factors that may worsen sickling and pulmonary injury. Nigerian pediatric guidance also emphasizes prompt treatment rather than waiting for the exact cause of an episode to be fully established (Adegoke et al., 2024).

Oxygen therapy is required when oxygen saturation falls below the patient's usual level or clear hypoxemia is present. Correcting hypoxemia is important because low oxygen tension can promote further sickling. Mild and moderate cases may respond to oxygen delivered through a nasal cannula or face mask, while patients with increasing oxygen requirements need closer observation. Persistent hypoxemia, greater work of breathing, or worsening mental status may indicate the need for non-invasive respiratory support or mechanical ventilation.

Antibiotics are commonly started early because infection cannot always be distinguished from ACS at presentation. Fever, cough, leukocytosis, pulmonary infiltrates, and hypoxemia may occur in both pneumonia and ACS, and infection itself is a recognized trigger. Vichinsky et al. (2000) identified bacterial, viral, and atypical pathogens in patients with ACS, although a definite infectious cause was not established in every episode. Empirical treatment should therefore cover common respiratory pathogens, with later adjustment according to microbiological findings, local resistance patterns, and clinical response.

Pain control also requires balance. Severe chest, back, or abdominal pain can limit deep breathing and promote atelectasis, so adequate analgesia is necessary. Opioids may be needed in significant vaso-occlusive pain, but excessive sedation can suppress ventilation. Patients receiving stronger analgesics should therefore be observed for changes in respiratory rate, oxygen saturation, and level of consciousness. Fluid therapy should also be cautious. Dehydration should be corrected, but excessive intravenous fluid may worsen pulmonary congestion. The goal is adequate hydration rather than routine aggressive fluid loading.

Measures that improve lung expansion can be useful, particularly when pain causes shallow breathing. Incentive spirometry has been shown to reduce pulmonary complications in hospitalized patients with sickle cell disease (Bellet et al., 1995). The Nigerian pediatric guideline also suggests practical alternatives, including repeated balloon blowing when a conventional spirometer is unavailable (Adegoke et al., 2024). Bronchodilators are mainly useful when wheezing, bronchospasm, or underlying asthma is present and are not required routinely in every ACS episode.

Blood transfusion becomes particularly important when ACS is associated with worsening anemia, persistent hypoxemia, or progression of pulmonary involvement. Simple transfusion can improve oxygen-carrying capacity and increase the proportion of normal red cells. It is most useful when hemoglobin has fallen substantially below the patient's baseline and the clinical condition is deteriorating. Transfusion should not simply aim to normalize hemoglobin, since excessive increases in hematocrit may increase blood viscosity in SCD. The decision should therefore be based on both the hemoglobin level and the overall clinical picture.

Exchange transfusion is generally reserved for more severe or rapidly progressive disease. By removing HbS-containing red cells and replacing them with donor cells, red cell exchange can reduce the HbS fraction more quickly while limiting the rise in blood viscosity. The American Society of Hematology suggests red cell exchange for severe ACS and considers it particularly relevant when the condition progresses rapidly, when simple transfusion produces an inadequate response, or when the baseline hemoglobin is already high enough to make further simple

transfusion less suitable (Chou et al., 2020).

Recent experience from Lagos shows that automated red cell exchange can be performed successfully in Nigeria. Adelekan-Popoola et al. (2026) reviewed 617 exchange procedures performed between 2020 and 2025, of which 74 were carried out for ACS. Across the full group, mean HbS fell markedly after treatment. The study is useful because it demonstrates that advanced transfusion therapy is feasible in a resource-constrained Nigerian setting. At the same time, access remains limited by equipment, trained personnel, blood supply, maintenance costs, and the concentration of specialist services in a small number of centers.

Respiratory support becomes necessary when oxygen therapy alone is no longer sufficient. Patients with persistent hypoxemia, increasing respiratory effort, acidosis, exhaustion, or altered consciousness should be assessed early for intensive care. Non-invasive ventilation may be useful in selected cases, but it should not delay intubation when respiratory failure is clearly progressing. In the National Acute Chest Syndrome Study, a proportion of patients required mechanical ventilation, showing that severe ACS can progress beyond the point where standard ward-level support is adequate (Vichinsky et al., 2000).

The Lagos case reported by Alli et al. (2012) also illustrates the need for repeated reassessment. Their 28-year-old patient initially received antibiotics, intravenous fluids, analgesia, and transfusion, but developed severe hypoxemia about 72 hours later and required intensive care support. The case shows that management cannot remain unchanged after the first treatment decision. When oxygen requirement rises, pulmonary infiltrates spread, or respiratory effort increases, treatment intensity needs to be adjusted quickly.

6. Challenges and Improvement of Management in Nigeria

The management of acute chest syndrome in Nigeria is affected by differences in access to diagnostic and treatment resources. Some patients are treated in tertiary hospitals with specialist hematology services, blood banks, imaging, and intensive care, while others first present to facilities with more limited support. This matters because ACS may initially resemble pneumonia or a vaso-occlusive pain crisis. Delayed recognition can reduce the time available for oxygen therapy, transfusion, or referral before severe hypoxemia develops. Nigerian multicenter data have shown that ACS is an important cause of respiratory-related mortality among hospitalized children and adolescents with SCD, which highlights the need for repeated respiratory assessment during the early stage of admission (Ibraheem et al., 2024).

Blood availability and access to exchange transfusion are also important limitations. Simple transfusion can be provided in many hospitals, but automated red cell exchange requires specialized equipment, trained personnel, compatible donor blood, and laboratory support. A five-year experience from Lagos showed that red cell exchange can be performed successfully in Nigeria and was used for ACS in a number of patients, but the program also faced difficulties related to cost, equipment maintenance, staffing, consumables, and blood supply (Adelekan-Popoola et al., 2026). For severe ACS, clearer referral pathways between general hospitals and specialist centers could help reduce delays in access to advanced transfusion and respiratory support.

Improvement should therefore focus on strengthening basic care at the point where patients first present. Hospitals treating patients with SCD should be able to recognize ACS early, monitor oxygen saturation, provide oxygen and appropriate supportive treatment, and identify signs that require urgent referral. The Paediatric Association of Nigeria guideline provides a useful local framework and adapts some recommendations to available resources (Adegoke et al., 2024). Long-term SCD management is also important. Regular follow-up, appropriate use of hydroxyurea, vaccination, infection control, asthma management, and patient education may reduce the risk of recurrent acute complications.

The main goal is not to make every hospital provide the same level of specialist treatment. A more practical approach is to ensure that basic recognition and stabilization are available widely, while severe cases can be transferred quickly to centers with transfusion, intensive care, and apheresis capacity. This would make the management of ACS more consistent without depending on advanced resources at every level of the health system.

References

Adegoke, S. A., Adeodu, O. O., & Adekile, A. D.

(2015). Sickle cell disease clinical phenotypes in children from South-Western, Nigeria. *Nigerian Journal of Clinical Practice, 18*(1), 95–101. https://doi.org/10.4103/1119-3077.146987

Adegoke, S., Adeola, S., Ahmad, H., Ambe, J., Dawodu, O., Diaku-Akinwumi, I., Ekure, E., Garba, M., Ibrahim, U., Israel-Aina, Y., James, E., Mohammed-Nafi'u, R., Nnebe-Agumadu, U., Odunvbun, M., Ofakunrin, A. O., Ogundeyi, M., Ogunrinde, O. G., Okechukwu, C., Okpe, E., ... Udechukwu, N. (2024). Paediatric Association of Nigeria guidelines on the management of acute chest syndrome in children with sickle cell disease (2023). *Nigerian Journal of Paediatrics, 51*(1), 37–54. https://doi.org/10.4314/njp.v51i1.07

Adelekan-Popoola, F., Gold-Olufadi, S., Kalejaiye, O. O., Ogala, A., & Balogun, R. A. (2026). Establishing an apheresis medicine program in a resource-constrained setting: A 5-year experience from Lagos, Nigeria. *Therapeutic Apheresis and Dialysis*, 1–7. https://doi.org/10.1002/1744-9987.70176

Ahmed, S. G. (2011). The role of infection in the pathogenesis of vaso-occlusive crisis in patients with sickle cell disease. *Mediterranean Journal of Hematology and Infectious Diseases, 3*(1), e2011028. https://doi.org/10.4084/MJHID.2011.028

Alli, F. O., Adebayo, A. K., Olowojolu, O. P., Daini, O. M., Osunkoya, S. A., & Thomas, B. O. (2012). Feasibility of early diagnosis and treatment of acute chest syndrome in sickle cell anaemia—A case report. *Nigerian Journal of Medicine, 21*(4), 469–471.

Bellet, P. S., Kalinyak, K. A., Shukla, R., Gelfand, M. J., & Rucknagel, D. L. (1995). Incentive spirometry to prevent acute pulmonary complications in sickle cell diseases. *The New England Journal of Medicine, 333*(11), 699–703. https://doi.org/10.1056/NEJM199509143331104

Chou, S. T., Alsawas, M., Fasano, R. M., Field, J. J., Hendrickson, J. E., Howard, J., Kameka, M., Kwiatkowski, J. L., Pirenne, F., Shi, P. A., Stowell, S. R., Thein, S. L., Westhoff, C. M., Wong, T. E., & Akl, E. A. (2020). American Society of Hematology 2020 guidelines for sickle cell disease: Transfusion support. *Blood Advances, 4*(2), 327–355. https://doi.org/10.1182/bloodadvances.2019001143

Howard, J., Hart, N., Roberts-Harewood, M., Cummins, M., Awogbade, M., & Davis, B. (2015). Guideline on the management of acute chest syndrome in sickle cell disease. *British Journal of Haematology, 169*(4), 492–505. https://doi.org/10.1111/bjh.13348

Ibraheem, R. M., Abdulkadir, M. B., Aliu, R., Issa, A., Ibrahim, O. R., Bello, A. O., Abubakar, F. I., Oloyede, I. P., Olasinde, Y. T., Briggs, D. C., Bashir, M. F., Salau, Q. O., Garba, B. I., Ameen, H. A., Suleiman, M. B., Bewaji, T. O., & Shina, H. K. (2024). Burden and outcome of respiratory morbidities among children and adolescents with sickle cell disease—A retrospective review of emergency presentations in some Nigerian tertiary institutions. *PLOS ONE, 19*(5), e0303323. https://doi.org/10.1371/journal.pone.0303323

Vichinsky, E. P., Styles, L. A., Colangelo, L. H., Wright, E. C., Castro, O., & Nickerson, B. (1997). Acute chest syndrome in sickle cell disease: Clinical presentation and course. *Blood, 89*(5), 1787–1792. https://doi.org/10.1182/blood.V89.5.1787

Vichinsky, E. P., Neumayr, L. D., Earles, A. N., Williams, R., Lennette, E. T., Dean, D., Nickerson, B., Orringer, E., McKie, V., Bellevue, R., Daeschner, C., & Manci, E. A. (2000). Causes and outcomes of the acute chest syndrome in sickle cell disease. *The New England Journal of Medicine, 342*(25), 1855–1865. https://doi.org/10.1056/NEJM200006223422502