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# Pneumonia as a Sign of Sickle Cell Disease: About a Case with Review of the Literature

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## Abstract

**Background:** Pulmonary infections including pneumonia during sickle cell disease are common and fall within the definition of acute chest syndrome. This clinical syndrome is defined by the association of a recent thoracic radiological abnormality with a clinical manifestation: chest pain, fever, or dyspnea. **Methods:** This is an observational, descriptive study of a special case discovered during a pediatric consultation at the university clinics of Kinshasa. Its presentation being a case not yet found at the university clinics makes its publication valuable. **Result:** We report the observation of an 8-year-old patient with hand-foot-mouth syndrome and 2 previous pneumonias, without any notion of parental consanguinity. On his physical examination, he presented crackling rales at the base of the right lung, $O_2$ desaturation at 88% in open air. The emergency assessment revealed a hemoglobin level of 8 g/dl, a leukocytosis of 18,000/mm$^3$, a leukocyte formula: N83%L12% M5%E0%B0%; C Reactive Protein at 48 mg/l. The thick film was negative. The chest X-ray (Front) revealed right basal lung disease of presumably infectious origin. **Conclusion:** This work emphasizes the frequency of the occurrence of pneumonia and its potential seriousness in a little-known sickle cell patient.

## Subject Areas

Infectious Diseases

## Keywords

Pneumonia, Sickle Cell Disease, Child, Kinshasa University Clinic

## 1. Introduction

Sickle cell disease is an autosomal recessive genetic hemoglobinopathy [1]. It is due to the substitution on chromosome 11 of the $\beta$ globin (HBB) gene of an amino acid in position 6 on the $\beta$ chain (HBB; c.20A > T, p.Glu7Val) [2]. The mutation of this gene located on chromosome 11 induces the synthesis of an abnormal hemoglobin (Hb), HbS, whose polymerized structure when deoxygenated profoundly modifies the properties of erythrocytes [1]-[3]. This results in hemolytic anemia which is the most common cause [2] [4]. Hypoxia is the main player in sickle cell disease. Acute, it initiates the cascade of physiopathological mechanisms which lead in the long term to chronic hypoxia. All lung compartments can be affected. Current treatments allow a life expectancy of more than 50 years [5] [6], but it remains in low-income countries responsible for significant mortality among children [4] [7]. There are different haplotypic clusters of $\beta$ globin, due to mutations occurring independently: four mutations of African origin (Senegal, Benin, Bantu and Cameroon) and one Asian, Arab-Indian (eastern province of Saudi Arabia and central India), the severity of which varies [4] [7]. Non-African mutations give clinical forms of less severity. In addition, polymorphisms in other genes influence the severity of the manifestations, the best known being those associated with the persistence of high levels of fetal hemoglobin (HbF, $\alpha 2\gamma 2$), found in the less severe phenotypes. Polymorphisms of BCL11-A, HBS1L-MYB and HBB modify the HbF level and are correlated with the frequency of painful crises, acute chest syndromes and the risk of infection [4] [8] [9].

Located in the heart of Africa, the Democratic Republic of Congo (DRC) is one of the countries most affected by sickle cell disease with 25 to 30% of Congolese carrying the S gene [10] [11]. During their life, children with sickle cell disease are likely to develop functional asplenia making them vulnerable to encapsulated germs, leading to potentially serious infections including those of the lower airways [12]. Knowing how to diagnose and manage respiratory infections in sickle cell patients will help improve their vital prognosis. It is in this context that the present case is documented.

## 2. Methods

A literature search by Pubmed, Google Scholar was conducted to strengthen in detail the case observation and the data published in the literature. The case was received at the University Clinics of Kinshasa during the general pediatric consultation. We described the sociodemographic characteristics of the patient, his clinic on admission and his paraclinical findings. We therefore made a case report described in the observation**.** The case was observed respecting the 3 principles of research ethics on beings, confidentiality was too rough and the parent had verbally consented that the case be published.

## 3. Observation or Result

This is the little girl Bwa, aged 8 years, 2nd of a family of 3 children, regularly

vaccinated, transfused once at the age of 8 months, with the notion of hand-foot-mouth syndrome and 2 previous cases of pneumonia, without the notion of parental consanguinity. She was taken to a consultation for a persistent cough and fever for 11 days for which the mother had consulted several health facilities, and where the little girl had been treated with Amoxicillin, Erythromycin, and other products of an unspecified nature without success. The persistence of the symptoms will motivate consultation at the University Clinics of Kinshasa (CUK) for better care. On his physical examination, he was mainly noted to have an unwell appearance, feverish at 39.4˚C, moderately colored palpebral conjunctivas, anicteric bulbar conjunctivas, tachycardia at 132 bpm, polygenic at 40 cpm, with intercostal drawing, an increase in the transmission of vocal vibrations, a decrease in gallbladder murmur, with crackling rales at the base of the right lung, $O_2$ desaturation at 88% in the open air, and he was also noted to have splenomegaly 4 cm below the costal margin LEFT. His neurological examination was normal. The hypothesis of left basal pneumonia having been raised, the emergency assessment revealed a hemoglobin level of 8 g/dl, a leukocytosis of 18,000/mm$^3$, a leukocyte formula: N83%L12% M5%E0% B0%; C Reactive Protein at 48 mg/l. The thick film was negative. The chest x-ray (Front) revealed right basal lung disease of presumably infectious origin. (See **Figure 1**)

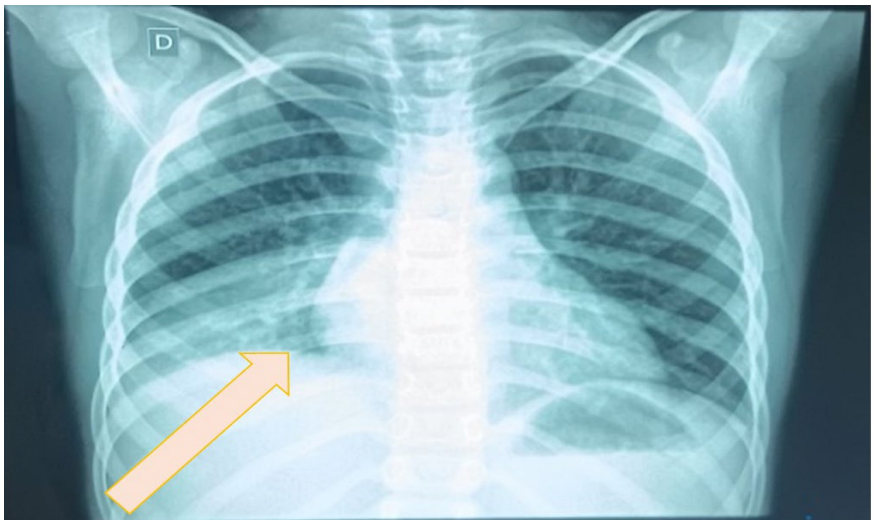


**Figure 1.** Image showing acute right basal lobar frank pneumonia revealing sickle cell disease in an 8-year-old girl. Ngonde, CUK, 2024.

In view of his personal history, a molecular test for sickle cell anemia was requested and carried out at the Faculty of Medicine of the University of Kinshasa. The said test revealed a homozygous sickle cell (SS) profile. The little girl was hospitalized for a total stay of 15 days, treated mainly with Augmentin-clavulanic acid/Amikacin, 48 hours of oxygen therapy, antipyretics for each febrile outbreak.

Having progressed well, she left the hospital with a recommendation to take daily penicillin V, folic acid, sufficient hydration and a follow-up program in the hematology department of CUK.

## 4. Discussion

Due to their condition, people with sickle cell disease are at high risk of developing an acute infection of the lung parenchyma [13]. It was the occurrence of pneumonia with severe respiratory distress that motivated the mother to consult and accept the molecular test for sickle cell anemia through our initiative. Many children die without diagnosis either because of financial barriers to diagnostic means, or because of the lack of recognition of the signs by caregivers and parents of the clinical picture of the disease. The absence of a national care policy, like most African countries, delays early intervention in the face of problems that can be avoided through vaccination therapy, antibiotics and folic acid supplementation, exposing sickle cell patients to complications. sometimes life-threatening such as pulmonary infections [11] [14]. It should first be mentioned that 50% of pulmonary infections develop in patients who initially presented with a painful attack alone, with an initially normal chest x-ray. The main symptoms are fever, cough, dyspnea and chest pain [15]. As hypoxemia is the main stimulus for polymerization, which itself leads to occlusion of the capillaries, even minimal initial pulmonary damage can degenerate into respiratory distress, then into Acute Respiratory Distress Syndrome with its dramatic consequences [16]. In a prospective series of 538 patients, 13% of patients with pneumonia required a diagnosis of sickle cell disease [15]. However, it is currently difficult to determine the proportion of patients with pneumonia who are sickle cell patients; in fact, this depends on various elements such as the clinical presentation of the patients, the history and the assessment carried out. It should be noted that pulmonary complications induced by pneumonia aggravate the thoracic syndrome and can be a direct sign of the onset of sickle cell disease [17].

## 5. Conclusion

Sickle cell disease remains a major public health problem in our environment. Systematic early detection of the disease, neonatal at best, could help avoid late discoveries whose consequences can be fatal.

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## Authors' Contributions

FNN conceptualized the research topic, ANN, CB, JMK, PMNS drafted the protocol with input from GNN for the methods, prepared the submission for institutional review board approval, supervised the data collection and drafted the manuscript. LPA provided content oversight for the manuscript. All authors

read and approved the final manuscript.

## Availability of Data and Materials

The datasets analyzed during this study are available from corresponding author on reasonable request.

## Conflicts of Interest

The authors declare no known conflict of interest.

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