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Acute thoracic syndrome in children with sickle cell disease hospitalized in the paediatric department of the University Hospital Gabriel Touré (UH-GT)

Abstract

The aim of our work was to study the epidemiological and clinical aspects of acute thoracic syndrome in children with sickle cell disease hospitalized in the paediatric department of the Gabriel Touré University.

Materials and methods: We conducted a retrospective study over six (06) years, from January 1, 2017 to October 31, 2022, based on the records of children under 16 years of age with sickle cell disease confirmed by haemoglobin electrophoresis and hospitalized for acute thoracic syndrome in the paediatric department of the UH-GT.

Results: The frequency of hospitalization was 5.46%, the average age was 6.2 years, with a male predominance (sex ratio: 4.6). Nearly half of (studie subject?) (47.8%) had a history of vaso-occlusive crises. Chest pain was the main reason of consultation (61.5%). Hyperthermia, tachypnoea and hypoxia were present in 43.6%, 4.4% and 76.9% respectively. Pulmonary signs were dominated by respiratory distress (92.3%), with the homozygous form (SS) being the most common (92.3%). Bilateral basal opacities were found on frontal chest X-ray in 46.2% of patients. All patients were rehydrated. The mean duration of treatment was 9.1 days, with extremes of 2 and 31 days. Mortality rate was 2.6%, related to the degree of hypoxia (P= 0.001).

Conclusion: Acute thoracic syndrome is a frequent and serious acute complication of sickle cell disease. Any chest pain associated with respiratory signs and poor oxygen saturation should call for the diagnosis Acute thoracic syndrome.

Keywords: sickle cell disease, acute thoracic syndrome, paediatric department, Bamako

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Introduction

According to WHO, over 120 million people worldwide carry sickle cell trait, and around 500,000 children are born homozygous for SS every year.[1] Sickle cell disease is predominantly found in sub-Saharan Africa with prevalence ranging from 15% to 45%.[2]

In Mali, around 12% of the population carries sickle cell trait, and 5,000 to 6,000 children are born with a major sickle cell phenotype every year.[3] Subjects with homozygous sickle cell trait SS or associated with another hemoglobinopathy (double heterozygous sickle cell trait S/C, S/β-thalassemia) suffer from complications of the disease.[4]

The pulmonary complications that punctuate the course of sickle cell disease are life-threatening for children in resource-limited countries.

Acute thoracic syndrome (ATS) represents a specific form of pulmonary aggression that can evolve into "acute respiratory distress syndrome". In 2009, a new definition of ATS was proposed: a clinical picture combining radiological evidence of a new pulmonary infiltrate with fever, desaturation or respiratory signs.[4]

ATS is one of the most frequent complications of sickle cell disease with a higher incidence in young children (12.8 episodes per 100 patient years).[5] It is the second most common reason for hospitalization and is responsible for 25% of premature deaths. Its occurrence is directly correlated with circulating leukocyte and hemoglobin levels, and inversely with age and hemoglobin F levels. Furthermore, patients with ATS have a higher risk of repeated vaso-occlusive crisis and are at risk of developing chronic obstructive pulmonary disease.[6] Although the Department of Pediatrics of the UH-GT has a unit for the care and follow-up of children with sickle cell disease, no study has focused specifically on thoracic syndrome in children. Therefore, we have initiated this work to contribute to a better understanding of APS in children and to improve cases management in our context.

Objectives: To study the epidemiological and clinical aspects of ATS in children with sickle cell disease hospitalized in the pediatrics department of the UH-GT.

Patients and methods

This was a retrospective study over six (06) years, from January 1, 2017 to October 31, 2022. It concerned the records of children under 16 years of age followed in the pediatric department of UH-GT for sickle cell disease confirmed by hemoglobin electrophoresis and presenting an ATS. Acute thoracic syndrome was defined as any child with sickle cell disease presenting a respiratory distress associated with one or more of the following symptoms: chest pain, fever, and cough. Data were collected on a dedicated survey form. The variables studied were epidemiological (age, sex, education level, residence, parents consanguineous marriage, etc); clinical (reason for consultation/referral, history, clinical signs); biological (CBC, reticulocyte count, hemoglobin electrophoresis, Rhesus group and thick drop); Radiological (Front and side thoracic radiographs, lung ultrasound, thoracic CT scan); therapeutic (hydration, antibiotic therapy, evolution).

Data were collected, entered and analyzed using SPSS version 22 software.

We used the chi-square test to compare qualitative variables. A value of $p<0.05$ was considered statistically significant. Data were collected anonymously.

Results

Epidemiological characteristics

Among, the 714 sickle-cell-disease affected children followed up in the pediatric department of UH-GT, 39 cases presented with acute thoracic syndrome, giving a frequency of 5.46%. This included 32 boys (82.1%) and 7 girls (17.9%), with an average age of 6.2 years and extremes of 06 months and 15 years. The age group, 6-10 years was the most represented (48.7%). (Table 1)

Table 1 Epidemiological characteristics

Characteristics		Effective (n=39)	%
Age (years)	Mean	6,2	
	6 months-5	16	41
	6-10	19	48,7
	11-15	04	10 ,2
Sex	Male	32	82,1
	Female	07	17,9

Clinical features

Nearly half of subjects with thoracique syndrome (47.8%) had a history of vaso-occlusive crises. In 74.4% of cases, sickle cell disease was discovered between the ages of 6 months and 5 years, and 56.4% were regularly monitored. The average time delay to consultation was 5 days. Chest pain was the main reason for consultation (61.5%). Hyperthermia, tachypnea and hypoxia were present in 43.6%, 4.4% and 76.9% respectively. (Table 2)

Table 2 Patients distribution according to baseline vital signs

Entry constants	Effective (n= 39)	%
Temperature		
Normal	9	23,1
Fever	13	33,3
Hyperthermia	**17**	**43,6**
Respiratory frequency		
Normal	4	10,2
Tachypnea	**29**	**74,4**
Bradypnea	6	15,4
Oxygen saturation (%)		
30 – 50*	1	2,6
51 – 70	2	5,1
71 – 90	**27**	**69,2**
91 – 94	8	20,5
≥ 95	1	2,6

***=** chronic cough (2), respiratory discomfort (1).

Pulmonary signs were dominated by respiratory distress (92.3%), bilateral rales accounted for 53.8%, extra pulmonary signs were pallor (94.9%) and jaundice (30, 8%).The homozygous sickle cell disease phenotype (SS) was the most represented (92.3%). Hyperleukocytosis represented 97.3% and severe anemia 62.2%. Bilateral basal opacities were found in 46.2% of cases on frontal chest X-ray.

Therapeutic aspects

All patients were rehydrated with lactated Ringer's, isotonic saline and 5% glucose. All had received Paracetamol and Tramadol. Third-generation cephalosporins, aminoglycosides and glycopeptides were used in 84.6%, 53.8% and 53.8% respectively. Transfusion with packed red blood cells was used in 61.5% of cases. Oxygen therapy was used in 97.4% of patients, and respiratory physiotherapy in 35.9%.

Evolution

The duration of treatment was 6 to 10 days in 51.3% of cases, with a mean of 9.1 days and extremes ranging from 2 to 31 days. Mortality was 2.6% (Table 3), related to age at onset (p=0.04) (Table 4), oxygen saturation at onset (p=0.001) (Table 5), quality of follow-up (p=0.02) (Table 6), and time to consultation (p=0.02). (Table 7)

Table 3 Patients distribution by outcome

Outcome	Effective	%
Healed	35	89,7
With Sequellae		**7,7**
Deceased	**1**	**2,6**
Total	**39**	**100,0**

Table 4 Patients distribution by outcome and age of discovery

Outcome	Age of discovery			Total
	06 to 24months	02 to 05years	06 to 10 years	
Healed	9	16	10	35
With sequellae	1	**2**	0	3
Deceased	0	**1**	0	1
Total	10	**19**	10	39

X2= 1,5 ; ddl= 4 ; P= 0,04.

Table 5 Patients distribution by outcome and oxygen saturation

Oxygen saturation (%)	Outcome			Total
	Healed	With sequellae	Deceased	
30 to 50	0	0	**1**	1
51 to 70	2	0	0	2
71 to 90	26	**1**	0	27
91 to 94	6	**2**	0	8
> 94	1	0	0	1
Total	**35**	**3**	**1**	**39**

X^2= 59,6 ; ddl= 30 ; P= 0,001

Table 6 Patients distribution by appointment follow-up and outcome

Appointment follow-up	Outcome			Total
	Healed	With sequellae	Deceased	
Regular	22	0	0	**22**
Irregular	13	3	1	**17**
Total	35	3	1	**39**

X^2= 5,8 ; ddl= 2 ; P= 0,02.

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Table 7 distribution of patient outcome according to Delay to hospitalization

Delay to hospitalization (days)	Outcome			Total
	Healed	With sequellae	Deceased	
≤ 03	9	0	0	9
04 to 07	22	0	0	22
08 to 14	3	**3**	**1**	7
> 14	1	0	0	1
Total	35	**3**	**1**	39

X^2= 20,4 ; ddl= 6 ; P= 0,02

Discussion

From January 1, 2017 to October 31, 2022, we collected 39 cases of children presenting acute chest syndrome out of 714 sickle cell disease children followed up in the pediatric department of UH Gabriel Touré, representing a frequency of 5.46%. Boiro et al.[7] in Dakar recorded a lower frequency (2.9%), whereas Kolie[8] obtained a much higher result (66.7%). The sickle-cell disease unit in the pediatric department of the UH-GT is not the only structure in the capital, and our prevalence may be underestimated because there is a sickle-cell disease research and control center which receives all age categories for follow-up and treatment of complications. The mean age was 6.2 years, in line with those of several authors.[8,9] Indeed, it is around the age of 06 that HbS almost totally replaces HbF, leading to an increase in the frequency of attacks and complications. In our resource-limited countries, there is a delay in diagnosis of the disease from 6 months to 5 years, which would explain the discovery of the disease at the stage of complications. Boiro et al.[7] found that 39.9% of cases were discovered in infants. More than half of patients had a history of hospitalization (59.0%). In our study Vaso-occlusive crises were the reason for previous hospitalization in 47.8%. The same observation was made by other authors.[7,8,10] This shows that we must actively manage the factors that trigger vaso-occlusive crises, which are essentially infections.

The main reason for consultation was chest pain (61.5%). The same finding was made by Kolie et al.[8] (41.7%). Chest pain can be a trigger for a pulmonary complication or contemporaneous with it.

More than half of patients consulted us within 04 to 07 days (56.4%) after the beginning of symptoms. The average consultation time was 05 days. Douamba et al.[11] found an average consultation time of 10 ± 3.4 days, with a minimum of 2 days and a maximum of 90 days. Most of our patients' parents come from poor socio-economic backgrounds, forcing them to try self-medication with traditional medicines.

Three-quarters of patients had fever and tachypnea on admission. Jobert et al.[12] and Douamba et al.[11] reported similar results.

Nearly all (97.4%) of our patients had room air saturations below 95%. Bertholdt[9] found 50% of patients with saturation < 95%. The hypoventilation observed in acute thoracic syndrome is at the root of the poor oxygen saturation.

In the semiology of acute thoracic syndrome,and pulmonary signs predominated. So, we found respiratory distress (92.3%), dullness (66.7%) and crepitus rales (66.7%). Our frequencies are higher than those of Cissé[13] and Bertholdt,[9] who found 80% and 50% respiratory distress respectively.

We noted a predominance of the SS phenotype with 92.3%, followed by the SC and Sβ° thalassemia forms with 5.1% and 2.6%. This same finding has been made by some authors.[8,9,14,15] However, it is contrary to the study by Ayéroué et al.[16] in Ouagadougou, who recorded 62% SC versus 38% SS. The high prevalence of the SS phenotype in the Sahelian zone has been demonstrated by several studies.[9,14,15] Hyperleukocytosis was noted in 97.3% of our patients, with a mean of 28403/mm3 leukocytes and extremes of 10500 and 60600/mm3. Douamba et al.[11] found that 84.9% of patients had hyperleukocytosis, with a mean leukocyte count of 22945/mm3 and extremes of 6400 and 78600/mm3. Hyperleukocytosis in sickle cell patients is classic, can be deleterious in certain situations (very high white blood cell count) and is associated with the risk of death.

Hemoglobin levels below 10g/dl were found in 97.3% of patients, with a mean of 5.3g/dl and extremes of 3.6g/dl and 10.2g/dl. Our results are comparable to those of Douamba et al,[11] who found a mean hemoglobin level of 6.7 g/dl with extremes of 2.5g/dl and 10g/dl, but lower than those of Bertholdt,[9] who found a mean hemoglobin level of 8.3 g/dl with extremes of 7.3g/dl and 9.5g/dl. Anemia is a constant feature of sickle cell disease due to chronic hemolysis, which can worsen in acute situations.

Normocytosis was found in 48.7% of patients presenting with a vaso-occlusive crisis, with a mean of 81.5 fl and extremes of 62.7 fl and 102 fl. Bertholdt[9] found that 48% (n=12) of patients had normocytic anemia. This may be explained by the normocytic anemia that is a feature of sickle cell disease.

All our patients underwent frontal chest radiography, which revealed bilateral basal opacities in 46.2% of patients.

This result is lower than that of Douamba[11] and Cissé ME,[13] who found 76.5% and 64.4% abnormalities on chest radiography. This difference can be explained by the delay of imaging in relation to the clinic, and also a normal chest radiograph does not eliminate the diagnosis of acute chest syndrome.

In line with the literature,[11–13] treatment consisted of rehydration with Ringer lactate (RL) and isotonic saline (IS) solutions 0.9% in 92.3%, probabilistic antibiotic therapy with third-generation cephalosporins (C3G) (84.6%) followed by vancomycin and gentamycin (53.8% each), and level I and II analgesics in all patients (100%). The majority of our patients (94.9%) were placed on oxygen. Oxygen flow was less than 3L in 66.6% of cases (n=38) for 3-6 days in 70.3% of cases. Our results are superior to those of Nicole[13] and Berthold,[9] who used oxygen therapy in 68.9% and 50% of their patients respectively. This use of oxygen is explained by the desaturation frequently observed in the pulmonary complications of sickle cell disease.

Transfusion in sickle-cell patients requires the presence of phenotyped packed red blood cells, which are difficult to obtain in emergency situations. Over half of the patients (61.5%) had received unphenotyped packed red blood cells (n=29). Jobert[12] recorded 66.7% of transfusions with whole blood, whereas Berthold et al[9] transfused 44% of their patients with packed red blood cells.

Over a third of patients (35.9%) had benefited from respiratory physiotherapy. Ideally, respiratory spirometry should be performed in all patients over 5 years of age, but lack of financial resources and the unavailability of this test were among the difficulties encountered in our study.

Acute chest syndrome is one of the most serious complications of sickle cell disease, and its management requires hospitalization. The mean hospital time was 9.1 days, with extremes of from 2 and 31 days, comparable to the results of Douamba[11] and Jobert.[12]

We recorded one case of death (2.6%) in a context of cardiac malformation on homozygous sickle cell disease SS received in a picture of severe hypoxia with a SaO2 of 31% on room air. This result is lower than that of Douamba,[11] who reported 7.5% (n=10) cases of death in children with major sickle cell syndromes and associated infections in Burkina Faso. ATS is a dreadful complication of sickle cell disease, whose prognosis depends on the severity of the clinical picture and the promptness of treatment. Indeed, the deceased patient had consulted the hospital after a relatively long delay and was in poor general condition at admission.

We noted a significant relationship between outcome and age of onset (P= 0.04), between follow-up appointments and outcome (p=0.04), between outcome and oxygen saturation (P= 0.001), and between outcome and time to consultation (P= 0.02).

Conclusion

ATS is a frequent complication of major sickle cell disease in our setting; Adoption of a regular follow-up will help reduce its proportion.

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Conflicts of interest

The authors declare that they have no conflicts of interest.

References

1. WHO. *Sickle cell disease: a strategy for the WHO African region*. AFR/RC. 2010;60/8:9p.
2. Ebakisse-Badassou E. The International sickle cell organization (ISCO) and the fight against sickle cell disease. *Méd Trop*. 2010;70(5):464–466.
3. Diallo D A. Sickle cell disease in Africa: Issues, strategies for improved follow-up and quality of life in sickle cell disease. *Bull Acad Natle Med*. 2008;192(7):1361–1373.
4. Ballas S K, Lieff S, Benjamin L J, et al. Definitions of the phenotypic manifestations of sickle cell disease. *Am J Hematol*. 2010;85(1):6–13.
5. Minter K R, Gladwin M T. Pulmonary complications of sickle cell anemia. A need for increased recognition, treatment, and research. *Am J Respir Crit Care Med*. 2001;164(11):2016–2019.
6. Vance L D, Rodeghier M, Cohen R T, et al. Increased risk of severe vaso-occlusive episodes after initial acute chest syndrome in children with sickle cell anemia less than 4 years old: sleep and asthma cohort. *Am J Hematol*. 2015;90(5):371–375.
7. Boiro D, Gueye M, Thiongane A, et al. Sickle cell disease in children, clinical and evolutionary profiles about 138 cases followed at the Pediatrics Department of Abass Ndao Hospital in Dakar. *Med Afr Noire*. 2016;63(6):326–332.
8. Kolié O O, Bangoura MA, Camara E, et al. Epidemioclinical and Therapeutic Aspects of Sickle Cell Disease in Children at Donka National Hospital (Conakry): a Study of 182 Cases. *Health Sci Dis*. 2023;24(4):127–131.
9. Bertholdt S, Le P Q, Heijmans S, et al. Respiratory complications of sickle cell disease in children: Acute chest syndrome. *Rev Med Brux*. 2012;33:138–144.
10. Mbika Cardorelle A, Okoko A, Mouko A. Vaso-occlusive crises in children with sickle cell disease in Brazzaville drugs news. *Arch Pediatr*. 2010;17(3):294–302.
11. Douamba S, Nagalo K, Tamini L, et al. Major sickle cell syndromes and associated infections in children in Burkina Faso. *Pan Afr J Med*. 2017;26(7):1–6.
12. Nansseu JRN, Alima Yanda A N, Chelo D, et al. The acute chest syndrome in Cameroonian children living with sickle cell disease. *BMC Pediatrics*. 2015;15(131):1–8.
13. Cissé ME, Diakité AA, Dembélé A, et al. Pulmonary Complications in Children with Sickle Cell Disease Followed at the Pediatric Department of Gabriel Toure University Hospital. *Open J Pediatr*. 2022;12:89–99.
14. Diagne I, Ndiaye O, Moreira C, et al. Sickle cell syndromes in pediatrics in Dakar (Senegal). *Arch Pédiatr*. 2000;7(1):16–24.
15. Latoundji S, Anani L, Ablet E, et al. Sickle cell morbidity and mortality in Benin. *Med Afr Noire*. 1991;38:8–9.
16. Ayéroué J, Kafando E, L Kam, et al. Sickle cell syndrome of the hemoglobin SC type: experience at the Yalgado Ouédraogo University Hospital in Ouagadougou (Burkina Faso). *Arch Ped*. 2009;16(4):316–321.