Acute chest syndrome in sickle cell disease

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Acute chest syndrome (ACS) is defined as a new pulmonary infiltrate and some combination of fever, chest pain and symptoms and signs of pulmonary disease such as cough, dyspnea, and tachypnea. It is one of the most frequent complications in patients with sickle cell disease (SCD) causing significant morbidity and mortality. The frequency is variable reaching up to 45%. It accounts for more than 90% of hospital admission, and causes approximately 25% of death in patients with SCD. Despite its substantial morbidity and mortality, relatively little is known on the etiology and pathophysiology. Some cases are clearly due to infection, other causes include hypoventilation after opioid analgesics, splinting due to rib infarction and excessive intravenous hydration and fat embolism. The risk factors for development of ACS include homozygous sickle cell anemia (HbSS), younger age group, and lower hemoglobin (Hb) F and high steady state white blood cell count. There is a very limited information on this complication in children with SCD in the Northwestern Province of Kingdom of Saudi Arabia (KSA). This report demonstrated the clinical experience on ACS in children with SCD in Madina Region, Northwestern Province of KSA.

This is a retrospective study carried out at Madina Maternity and Children's Hospital (MMCH), Madina, KSA between January 1996 and January 2000. All pediatric patients (≤12 years) with SCD and developed ACS within the study period were included and case notes were reviewed. The following variable were studied: age at onset, sex, nationality, clinical presentation, hematological data at presentation, steady state hematological data, radiological finding on chest x-ray, management undertaken, recurrence and mortality. A total number of SCD patients registered and followed at MMCH during the study period were 120 of which 12 patients had ACS, which accounted for a prevalence of 10%. All patients had HbSS disease. The age range was between 2-13 years. They were 8 males and 4 females with a male to female ratio of 2:1. Nine were Saudis and 3 were non-Saudis. Eleven (92%) presented with fever, chest pain occurred in 6 [50%], 10 [83%] were associated with painful vaso-occlusive crisis and hypoxemia occurred in 2 [17%]. The ACS was recognized on presentation in 9 patients (75%) and was recognized later after admission in 3 (25%). The leukocyte count was more than 15,000/mm^3 in 8 patients (67%), and Hb was less than 6g/dl in 8 (67%). The chest x-rays showed right lower lobe involvement in 6 patients (50%), bilateral involvement in 4 (33%) and left lower lobe in 2 (17%). Bacterial cause was identified in 2 patients (17%) and undetermined in 10 (83%). Bronchial asthma was a precipitating factor in 5 (42%) patients. Eight patients (67%) received simple blood transfusion, recurrence occurred in 2 (17%), and there was no mortality. **Table 1** summarizes the hematological data during steady state.

This is the first study in the Madina Region to record the experience of ACS in children with SCD, and to the best of our knowledge, it has not been reported from the Western Province of KSA. In our study, the most affected age group was 5-10 years (50%), the youngest was 2 years, Al-Dabbous reported the youngest age affected was 9 months.[1] The majority of our patients was male (67%), which is consistent with other study.[1] The reason for male predominance is still unknown. All our patients were homozygous HbSS, high incidence of ACS in this type of SCD was also observed by others.[1] Fever was the most common presentation, (92%), cough and dyspnea were found in 75% of our patients and was the second common presentation after fever while chest pain was recognized in 50%. Painful vaso-occlusive crisis was associated with 83% of patients, it has been consider the most common associated event with ACS,[2] and high incidence of fat-embolism was the cause in ACS among those patients.[3] Hypoxia occurred in 2 patients (17%), this support the finding that ACS is mild in children in contrast to adult, as evidence by severe hypoxia, higher rate of transfusion, longer hospitalization and higher death rate.[2] Acute chest syndrome was recognized on presentation in 75% of our patients and was recognized later after admission

Table 1 - Steady state hematological data.

Age (year)	WBC x10^9/L	Hb g/dl	PLTs x10^9/L	Retic %	Hb electrophoresis HbA %	HbF %	HbS %	HbA2 %
12	16	6	300	7		20	78	2
07	10	7	450	4	40	-	56.4	3.6
07	16	8	460	5	18	20	60	2
09	16	7	316	5	15	12	70	3
04	12	7	560	7	10	20	66.5	3.5
10	20	7	450	7	-	30	67	3.1
11	10	9	360	8	15	20	65	-
12	10	8	200	8	-	10.6	85	3.4
13	16	10	145	4	10	15	75	-
02	20	7	400	6	27.4	-	70	2.6
09	18	8	550	10	40	-	58	2
08	17	6	360	8	17	-	80	3

WBC - white blood count, PLT -platelets, HbA - hemoglobin A, HbF - hemoglobin F, HbS -hemoglobin S, HbA2 - hemoglobin A2

in 25%, similar observation reported by other workers,[3] those patients usually presented with fever and painful crisis without respiratory symptoms or pulmonary infiltrate and ACS usually develops 2-3 days later. The radiological findings showed the lower lobes of the lungs predominantly involved, similar finding observed by other workers.[3] Bacterial infection was identified only in 2 patients (17%), one due to *Streptococcus pneumoniae* and other due to *Staphylococcus aureus*, low incidence of bacterial infection was also reported in other studies.[1] Most of the studies relied on blood cultures and is therefore likely to underestimate the frequency of bacterial pneumonia. Bronchoscopy is more sensitive method in determining the etiology of ACS. Despite low incidence of bacteremia in our patients all received empirical intravenous antibiotics (ceftriaxone + erythromycin), vancomycin was added for those with severe ACS and prolonged fever, considering the emerging resistance of pneumococcus to the usual antibiotics. The majority of our patients (67%) received simple blood transfusion (BT), and all show dramatic clinical improvement. Transfusion of packed red blood cells has a number of potential benefits, it increases oxygen carrying capacity, decreases the fraction of Hb S and thereby potentially reduces or reverses intrapulmonary sickling.[4] Simple BT should be given for marked decrease in Hb and also for clinical deterioration, however BT should be avoided in cases of Hb above 10g/dl to prevent increase blood viscosity, which may lead to stroke or exacerbate ACS. Exchange transfusion should be reserved for rapid clinical deterioration, widespread pulmonary involvement, hypoxemia not corrected by delivery of supplemental oxygen and multiorgan failure.[4] The low recurrence and death rate also support the observation of mild ACS in western region similar to the eastern province.[1] As the etiology of ACS appears to be multifactorial, there is no effective and specific therapy, although a beneficial effect of dexamethasone in children with mild to moderately severe acute chest syndrome was demonstrated in a prospective randomized double blind placebo controlled clinical trial,[5] the study showed significant reduction in the length of hospitalization, decrease duration of oxygen supplementation, decrease duration of opioid analgesia, decrease occurrence of clinical deterioration and decrease the need for BT. The steady state hematological data (**Table 1**) did not show clear risk factors for occurrence of ACS.

In conclusion, we found that the majority of ACS in our region is due to undetermined origin presumably secondary to rib infarction, atelectasis or fat embolism and further prospective study is required to confirm this possibility, we also conclude that bronchial asthma is an important precipitating factor for occurrence of ACS in SCD. Finally, we also recommend simple BT to be part of the management of ACS in children.

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Spectrum of childhood poisoning in a tertiary center in the Eastern Saudi Arabia

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Poisoning is a major and persistent cause of accidental morbidity in children worldwide despite being preventable. Fortunately, in most of the cases, toxic agent ingested has minimal significant effect. Although a lot of developments have been made in the prevention, diagnosis and treatment of pediatrics poisoning in the last 5 decades, yet accidental poisoning continues to burden the health care system worldwide and remains in one of the top 10 leading causes of death. There have been sparse data from the Kingdom of Saudi Arabia (KSA); therefore, we report a retrospective analysis of accidental poisoning in