Research Article

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Patterns and Presentation of Sickle Cell Disease in Children Attended Pediatrics Emergency Department-Security Forces Hospital-Riyadh-Saudi Arabia

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Introduction

Sickle cell disease (SCD) is a single gene disorder of human beings. In the United States, about 72 000 people had SCD, and 2 million are carriers [1]. In Africa, more than 200 000 infants per year are born with SCD [2]. In the 1960s, SCD was reported in Saudi Arabia [3]. Saudi Arabia started multiple regional and national screening studies to determine the clinical characteristics and frequency of SCD genes in different regions of Saudi Arabia [4-7]. The prevalence of SCD in Saudi Arabia is patchy, but now SCD is an almost common genetic disorder with a carrier status ranged from 2% to 27%, and up to 1.4% had SCD, in some areas [3-7].

According to the Saudi Premarital Screening Program, the prevalence of the sickle cell gene in the adult population is 4.2% for sickle-cell trait and 0.26% for SCD, with the highest prevalence noted in the Eastern province (approximately 17% for sickle-cell trait and 1.2% for SCD) [7].

In Saudi Arabia, sickle cell disease prevalence is 24 per 10,000, with a regional distribution that showed eastern region with a prevalence of 145 per 10,000, a southern region with a prevalence of 24 per 10,000, western region 12 per 10,000, and central region with 6 per 10,000 [8].

The SCD had unpredictable and variable clinical presentations. Recently, SCD clinical presentation was classified into two clinical phenotypes [9]. In the hemolysis-associated phenotype, the clinical presentation is severe anemia, leg ulcers, and pulmonary hypertension. In the vaso-occlusion-related phenotype, the clinical presentation is pain, acute chest syndrome, splenic infarction, stroke, and avascular necrosis of joints predominate. Endothelial dysfunction and vasculopathy are presented symptoms of SCD. Although the course of SCD is unpredictable, the disease is often associated with substantial morbidity, a decreased life span, and a poor quality of life. Early death is highest among patients who have had severe complications, such as recurrent acute chest

syndrome, renal failure, and pulmonary hypertension [10, 11].

Evidence and studies showed that early neonatal screening for SCD, early diagnostic testing, parental education, and comprehensive care markedly reduce SCD morbidity and mortality [12-14].

Methodology

The study was a retrospective cross-sectional study conducted in the Pediatrics emergency department at Security Forces Hospital-Al-Riyadh from 2013 to 2019.

Ethical approval was taken from the Institutional Ethical Review Board. All pediatrics patients with sickle cell disease aged up to 14 years were included in this study during the study period.

Excluded criteria were: those older than 12 years, Hb SC, Sickle Cell with Thalassemia, and those who attended ED before the year 2013 and after 2019. The details of the patients were taken from computerized medical records and patient files of the hospital during the study period.

Variables in the study included: demographic data (age and sex), year of attendance to ED, complain of the patient, blood culture result, diagnosis at presentation, and medication was given (like morphine). Data were analyzed by SPSS software version 24.

Result

This study was conducted in the emergency department of pediatrics at Security Forces hospital -Riyadh from 2013 to 2019. The total number of patients seen during the study period was 679 patients. Males were 483 patients (71.1%), and females were 196 patients (28.9%). The mean age was 6.826±3.1456.

In figure [1], age distribution showed: less than five years were 224 patients (33%), 5_10 year were 363 patients (53.5%), and more than ten years were 92 patients (13.5%).

Figure 1: Age distribution

In figure [2] the distribution of patients per year showed that in 2013 those seen ED were 93 patients (13.5%), 2014 were 88 patients (13%), 2015 were 71 patients (10.5%), 2016 were 117(17.2%), 2017 were (119(17.5%), 2018 were 127 patient (18.7%) and 2019 were 64(9.4%).

Figure 2: Distribution per year 2013 to 2019

Table [1] showed the diagnosis distribution: painful crisis in 447 patient (65.8%), hemolytic crisis in 8 patients (1.2%), acute chest syndrome (ACS) in 4 patients (0.6%), cholecystitis and gall stone in 3 patients (0.4%), aplastic crisis in 3(0.4%), infection in 175 patients (25.8%), sequestration crisis in 7 patients (1 %), and others (like otitis media, tonsillitis, gastroenteritis, etc..) in 32 patients (4.7%).

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Table 1 : Diagnosis

	Frequency	Per-cent	Valid Percent	Cumulative Percent
Valid Pain	447	65.8	65.8	65.8
Hemolytic crisis	8	1.2	1.2	67.0
ACS	4	.6	.6	67.6
Cholecystitis & Gall Stone	3	.4	.4	68.0
Aplastic Crisis	3	.4	.4	68.5
Infection	175	25.8	25.8	94.3
Sequestration Crisis	7	1.0	1.0	95.3
Others	32	4.7	4.7	100.0
Total	679	100.0	100.0	

Table [2] showed diagnosis and disposition correlation: admission was made to 24.4% of those with painful crisis, and 75.6% were discharged. 29.1% of those with Infection were admitted, where 70.9% with Infection and discharged. P-value was 0.000.

Table 2: Diagnosis & Disposition Cross tabulation p-Value=0.000

		Disposition		Total
		Admission	Dis-charge	
Diagnosis	Count	109	338	447
Painful crisis	% within Diagnosis	24.4%	75.6%	100.0%
	% of Total	16.1%	49.8%	65.8%
Hemolytic crisis	Count	8	0	8
	% within Diagnosis	100.0%	0.0%	100.0%
	% of Total	1.2%	0.0%	1.2%
ACS	Count	4	0	4
	% within Diagnosis	100.0%	0.0%	100.0%
	% of Total	0.6%	0.0%	0.6%
Cholecystitis & Gall Stone	Count	3	0	3
	% within Diagnosis	100.0%	0.0%	100.0%
	% of Total	0.4%	0.0%	0.4%
Aplastic Crisis	Count	3	0	3
	% within Diagnosis	100.0%	0.0%	100.0%
	% of Total	0.4%	0.0%	0.4%
Infection	Count	51	124	175
	% within Diagnosis	29.1%	70.9%	100.0%
	% of Total	7.5%	18.3%	25.8%
Sequestration Crisis	Count	7	0	7
	% within Diagnosis	100.0%	0.0%	100.0%
	% of Total	1.0%	0.0%	1.0%
Others	Count	10	22	32
	% within Diagnosis	31.3%	68.8%	100.0%
	% of Total	1.5%	3.2%	4.7%
Total	Count	195	484	679
	% within Diagnosis	28.7%	71.3%	100.0%
	% of Total	28.7%	71.3%	100.0%

In figure [3], morphine was given to 171 patients (25.2%).

Figure 3: Morphine

In figure [4] blood transfusion for 56 patients (8.2%)

Figure 4: blood transfusion..

Discussion

We studied 679 children seen in the pediatric emergency department of Security forces hospital from 2013 to 2019.

Most of our study group were between 5-10 years of age, representing 53.5%. The mean age of our study population was 6.8 ± 3.1 . Males were the dominant of patients attending the emergency department representing 71.1%. In Cameroonian children in Jobert Richie N Nansseu, et al. study male were the commonest who attended hospital, and their study is similar to our finding in the age group and mean age of study group [15].

Also, in Reem A. Hejazi, et al. study males patients who attended the emergency department are more than females [16].

Most of our patients seen in pediatrics emergency presented with painful crises, the second was infections, and other symptoms like otitis media, tonsillitis, and gastroenteritis represented the third common clinical presentation. Abeer A Abdel Moneim, et al. a study in the Western region of Saudi Arabia, was similar to our study result. It showed acute painful crisis as a common presenting complaint of admission; acute chest syndrome was reported in 20.9% and Infection in 17.5% of their patients [17].

Those with cholecystitis and gal stone were 0.4% in our study population, similar to a study done in the Democratic Republic of Congo that reported it as a rare presenting complication [18].

Morphine was given to about 25% of our patients attending the emergency department with pain; in Reem A. Hejazi, et al. study, most of their patient with painful crisis received morphine to control the pain [19].

Blood transfusion was done to 8.2% of our patients. In Yemen (77.9%) were transfused and received backed RBC transfusion episodes [20]. They had limited-resource settings, so conservative transfusion policy appears to be appropriate wherein our setting we were restricted to the policy and guidelines for blood transfusion.

The study showed a gradual increase in the number of patients seen per year from 2013 to 2019, and this due to improvement in services in the pediatrics emergency department.

Conclusion

Our study is similar to most of the studies that 5-10 years were the most common age group seen in the emergency department. Painful crises were the commonest presenting symptoms, followed by Infection as the second most common one. Most sickle cell disease children in the pediatrics emergency department with Infection were treated as an outpatient.

Conflicts of Interest: There are no conflicts of interest.

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