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Evaluation of Respiratory Function Test in Children with Sickle Cell Anemia with Acute Chest Syndrome

Mohammad Ali Molavi [1*] and Hamideh Doozandeh [2]

[1] *Department of Pediatric Hematology, Hormozgan University of Medical Sciences, Bandar Abbas, Iran*
[2] *Medical university of Hormozgan, Bandar Abbas, Iran*

* Corresponding author's Email: shayan molavi@yahoo.com

ABSTRACT: Sickle cell disease is caused by mutant recessive autosomal in β-globin chain leading to deformation of sickle cells. Hemoglobin S in deoxygenated conditions causes microvascular obstruction and adverse effects on various organs. The manifestation of sickle cell in lungs is as a thromboembolism, acute chest syndrome and abnormal lung activity test. This study aimed to Evaluation of the respiratory function test in children with sickle cell and its relationship with age, sex, allele type, acute chest syndrome and pain crisis, that lung injury of these patients to be diagnosed early. This descriptive-analytical study was done on 6-18 years old children in the Pediatric Hematology Clinic at Bandarabbas Hospital in 2011 to 2012. Following the CBC, Hb electrophoresis, spirometery and records of the patients (acute chest syndrome, pain crisis), theses date were recorded and imported to SPSS20 statistical software (mean ± SD) and analyzed by Chi-squar test. Of 29 patients, 17 were male and 12 were female. Highest genotype was SB 44.8 % (13 cases) and lowest was SA and SB0 6.9 % (2 cases). Results of lung function test were as follows: restrictive pattern (72.4%, 21 cases), normal pattern (20.7%, 6 cases) and obstructive pattern (6.9%, 2 cases). There was no significant difference in pattern of pulmonary function test in patients with and without history of previous acute chest syndrome. There was also no significant correlation between spirometric pattern and age, sex, type allele and pain crisis. Pulmonary function is abnormal in 79.3% children with sickle cell anemia. Common abnormalities include restrictive pattern.

Key words: Sickle Cell Anemia, Lung Activity Test, Acute Chest Syndrome

ORIGINAL ARTICLE

INTRODUCTIN

Sickle-cell anemia (SCD) is caused by a point mutation in the β-globin chain of hemoglobin, causing the hydrophilic amino acid glutamic acid to be replaced with the hydrophobic amino acid valine at the sixth position. SCD characterized by red blood cells that assume an abnormal, rigid, sickle shape. Sickling decreases the cells' flexibility and results in a risk of various complications. Sickle-cell anemia is the name of a specific form of sickle-cell disease in which there is homozygosity for the mutation that causes HbS. Sickle-cell anemia is also referred to as "HbSS", "SS disease", "hemoglobin S" or permutations thereof. In heterozygous people, who have only one sickle gene and one normal adult hemoglobin gene, it is referred to as "HbAS" or "sickle cell trait". Other, rarer forms of sickle-cell disease include sickle-hemoglobin C disease (HbSC), sickle beta-plus-thalassemia (HbS/β+) and sickle beta-zero-thalassemia (HbS/β0). These other forms of sickle-cell disease are compound heterozygous states in which the person has only one copy of the mutation that causes HbS and one copy of another abnormal hemoglobin allele. Life expectancy is shortened. In 1994, in the US, the average life expectancy of persons with this condition was estimated to be 42 years in males and 48 years in females (Platt et al., 1994) but today, thanks to better management of the disease, patients can live into their 50s or beyond (Rees et al., 2010). Sickle-cell disease may lead to various sign and symptoms (splenic sequestration crisis, Aplastic crisis, and Hemolytic crisis) and complications including: overwhelming post- (auto) splenectomy infection (OPSI), Stroke, Cholelithiasis, Priapism, Osteomyelitis, Pre-eclampsia, pulmonary hypertension, and Chronic renal failure. The acute chest syndrome (ACS) is an acute overwhelming pulmonary illness that occurs in patients with sickle cell disease. ACS is currently defined as a new infiltrate on chest radiograph in conjunction with 1 other new symptom or sign: chest pain, cough, wheezing, tachypnea, and/or fever (> 38.5°C) (Platt et al., 2000) The term acute chest syndrome was first suggested by Charache et al. (1979) and was developed to reflect the unique nature of acute pulmonary illness in patients with sickle cell disease. ACS can be caused by a variety of mechanisms, both infectious and noninfectious. Diagnostic considerations and treatment modalities are not typical of any other specific pulmonary illness experienced by the general

population. Furthermore, the typical course, possible complications, and outcomes are unique. For these reasons, the terminology persists and remains useful for both research purposes and effective clinical communication Due to the adaptive advantage of the heterozygote, the disease is still prevalent, especially among people with recent ancestry in malaria-stricken areas, such as Africa, the Mediterranean, India and the Middle East(Kwiatkowski., 2005) Malaria was historically endemic to southern Europe, but it was declared eradicated in the mid-20th century, with the exception of rare sporadic cases(Ponçon et al., 2007). The highest frequency of sickle cell disease is found in tropical regions, particularly sub-Saharan Africa, India and the Middle-East (Weatherall et al., 2001). Migration of substantial populations from these high prevalence areas to low prevalence countries in Europe has dramatically increased in recent decades and in some European countries sickle cell disease has now overtaken more familiar genetic conditions such as hemophilia and cystic fibrosis (Roberts et al., 2007). About 90% of patients survive to age 20, and close to 50% survive beyond the fifth decade (Kumar et al., 20090. In 2001, according to one study, the estimated mean survival for sickle cell patients was 53 years old for men and 58 years old for women with homozygous SCD (Wierenga et al., 2001).

METHODS AND MATERIALS

This descriptive – analytic cross- sectional study was done in the Bandarabbas Pediatric Hematology Clinic in March 2011 to March 2012. The study was conducted on children with sickle cell anemia aged 6 to 18 years old who admitted to Bandar Abbas Pediatric Hospital. Diagnosis of anemia was done by specialty hematology pediatric through electrophoresis and cell blood count. The children under 6 years old and patients with history of bronchial asthma and Atopic dermatitis excluded from study. Necessary information Provide to children with respect to children level of understanding and ability and attracting child's attention for cooperation. Informed consent was obtained from parents of children in the study. The data collected through lung functional test by experienced technician. Patient's records were reviewed and acute chest syndrome vascular occlusive crisis in their history were recorded. The data were analyzed by SPSS20 statistical software. P.value <0.05 was considered as significance level.

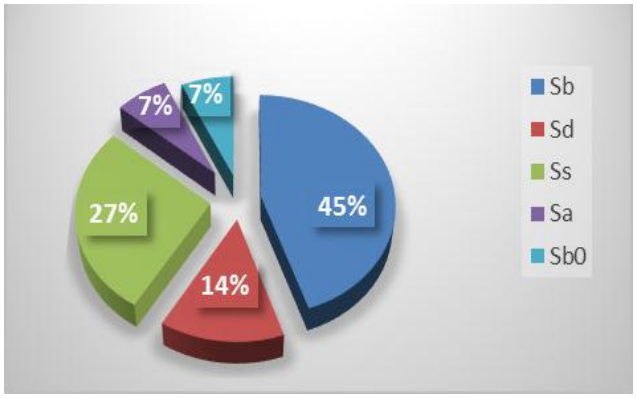
RESULTS

The average age of children was 10.17 with standard deviation 2.79. The most age group were 6 to 10 years old (55.2%) (Table 1). Frequency Distribution of alleles was shown in scheme 1. The most frequency was Sb allele (44.8%). Table 2 shows the Frequency Distribution of severity of disease in based on spirometric pattern. The most common spirometric pattern was restrictive pattern (72.4%) (Scheme 2). Spirometric patterns were examined in patients with or without acute chest syndrome (Table 3). There was no significant correlation between Spirometric patterns and acute chest syndrome according to table 3. In the base of FEV1 (ERS) only 3.4% of patients had severe pulmonary disease (Scheme 3). Spirometric patterns in different allele type were shown in Table 4.

There was no significant correlation between pulmonary patterns and allele type.Comparsion of pulmonary patterns in different age group was shown in table 5. The data in this table showed that there was no significant correlation between spirometric pattern and age. Table 6 showed that there was no significant correlation between spirometric pattern and patient's gender.

Table 1. Frequency Distribution of age

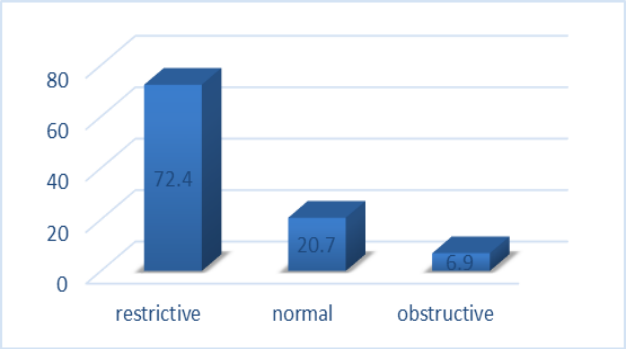
Age group(Year)	Frequency (%)	The average of age (Year)
6-10	55.2	8.06
10-14	37.9	12.27
14-18	6.9	15.50
Total	100	10.17



Scheme 1. The Frequency Distribution of alleles

Table 2. Frequency Distribution of severity of disease in based on spirometric pattern

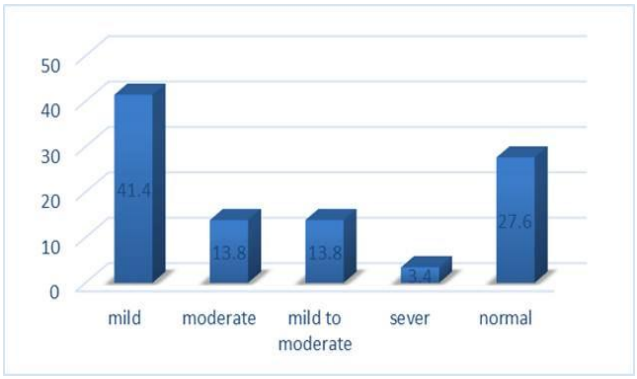
	Spirometry	Frequency (%)
Restrictive	Mild	57.1
	moderate	19
	moderate to severe	14.3
	Severe	4.8
Obstructive	Mild	-
	moderate	-
	moderate to severe	50
	Severe	-



Scheme 2. The Frequency Distribution of spirometric patterns

Table 3. The distribution of spirometric patterns in the base of with or without acute chest syndrome

Acute chest syndrome	Spirometry (%)			P. value
	Normal	Restrictive	Obstructive	
Yes	18.2	63.4	18.2	0.172
No	22.2	77.8	-	



Scheme 3. Frequency distribution of severity of pulmonary disease

Table 4. The distribution of spirometric patterns in the base allele type

Allele type	Spirometry (%)			P.*value*
	Normal	Restrictive	Obstruction	
Sb	7.7	84.6	7.7	0.064
Sd	75	-	35	
SS	12.5	87.5	-	
Sa	-	100	-	
Sb0	50	50	-	

Table 5. The distribution of spirometric patterns in different age group

Age group (Year)	Spirometry (%)			P.value
	Normal	Restrictive	Obstructive	
6-10	31.3	68.8	-	0.239
10-14	9.1	72.2	18.2	
14-18	-	100	-	

Table 6. The distribution of spirometric patterns in different gender

Gender	Spirometry (%)			P.value
	Normal	Restrictive	Obstructive	
Male	23.5	76.5	-	0.214
Female	16.7	66.7	16.7	

DISCUSSION

A total of 29 patients with sickle cell anemia who enrolled the study were 17 males (58.6%) and 12 females (41.4%) during one year in this study. pulmonary pattern was normal in 6 cases (20.7%) and abnormal in 23 cases (79.3%). The most common pulmonary pattern in this patients was restrictive pattern (72.4%) which consistent with studies carried by Klings ES (2006), Painosi (1993), Delclaux (2005) and Hulke (2011). Regarding to small blood sample volume, there was a significant difference. Such that more than half of volunteers were male (58.6%) and half of them had Sb allele (44.8%). In our study, there was no significant correlation between spirometric pattern and acute chest syndrome just like Elizabeth's and Painsoli's study but in contrast to Hlke's study. Restrictive pattern was the most common spirometric pattern in our study wherease the most spirometric pattern was normal in Hulke's study. The reason of this difference can be small blood sample volume (29 cases in our study, 133 cases in Hulke's study). But in large blood sample volume (Elizabeth's study), the most spirometric pattern was restrictive pattern (74%) similar to our study. like the Elizabeth's and Hulke's study, there was no significant correlation between age and gender with spirometric pattern in our study. In our findings, there was no significant correlation between allele type and pulmonary pattern unlike the Elizabeth's study.

Finally, there was no significant correlation between crisis pain and spirometric pattern. The results of this study revealed that physicians and carers need to be more attentive to spirometric pattern in children with sickle cell anemia in the younger age.

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