Abstract ID#: 122123

CHART REVIEW STUDY FOR PATIENTS WITH SICKLE CELL DISEASE WITH CENTRAL NERVOUS SYSTEM CRISIS IN JAZAN – SAUDI ARABIA

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ABSTRACT

OBJECTIVES: Descriptive analysis of patients suffering from Sickle-Cell Disease (SCD) with CNS (Central Nervous System) crises, and the correlation of different variables as demographics, major complications, hematologic and genetic factors on the degree of CNS crisis, for predictor generation and better future management.

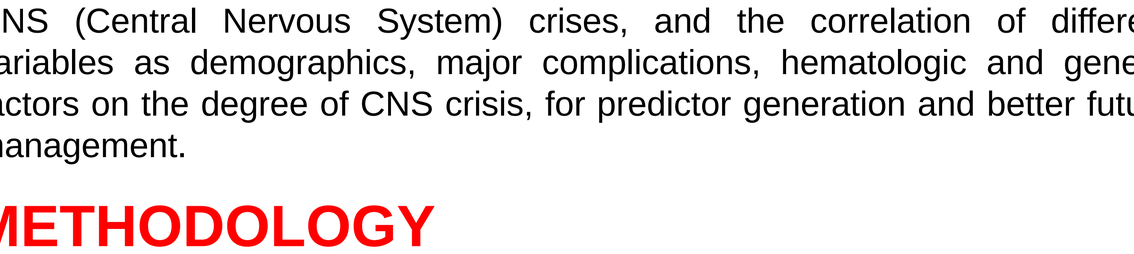
METHODS:A retrospective patients' chart review study was built with total of 146 SCD patients with CNS crisis inside the Hematology Department in Prince Mohamed Bin Nasser Hospital in Jazan (PMBNH) Saudi Arabia from Jan.1st to Dec. 31st, 2021. Analysis was done using SPSS for descriptive analysis and Jaccards for regression and multiple regression analysis.

RESULTS: Patients with SCD and CNS crisis were 72(49.3%) male and 74(50.7%) female with age range from 14 to 47 years and mean of 24.4. Most of them were single (76.7%) and non-employed (80.8%). Hemoglobin electrophoresis showed (73.3%) of patients to be homozygous SCA (HbSS), while (26.7%) were SC/β-thalassaemia (HbS/ßo and HbS/ß+). The frequency of CNS crisis was once in (89.0%), twice in (8.2%), and three times in (2.7%). The mean age at onset of first CNS crisis was 15.1 years. About (52.1%) developed the first CNS crisis before 10 years of age. The complications associated SCD were vaso-occlusive crisis (100%), avascular necrosis (16.4%), acute chest syndrome (7.5%), pulmonary hypertension and intrahepatic cholestasis (2.7% for each). The apparent CNS sequelae were found among (36.3%) of patients, included motor disability (60.4%), intellectual disability (32.1%), epilepsy (17.0%) and dysphasia (11.3%). About 70(47.9%) were successfully referred to higher centers for stem cell transplantation (SCT) and the case fatality rate within the study was 2.74%.

CONCLUSION:CNS crises are common morbidity among SCD patients, the optimized management of cases need close monitoring of CNS crises and follow-up for improved patients' outcome and avoiding further complications.

INTRODUCTION

Sickle cell disease is an autosomal recessive disease caused by a common single point mutation (valine instead of glutamic acid the sixth position).



Resulting in formation of hemoglobin S, a less soluble form of hemoglobin that once deoxygenated polymerizes to form abnormal crescent shape red blood cells. These cells are vulnerable to breakage and consequently causing hemolysis, vascular occlusion and ischemia.(1)

Sickle cell disease affected globally around 300,000 - 400,000 individual Per year

With High Mortality rate Among children in developing countries

In Saudi Arabia, the prevalence of SCD was highest in the Eastern province, followed by the southwestern provinces, mainly Jazan.(4) SCD is a common cause of morbidity and mortality in

OBJECTIVES

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METHODOLOGY

Retrospective Study For all SCD Patients Jan 1st to December 31st 2021

All SCD patients with CNS crisis attended the adult Hematology Department in PMBNH for follow up. The total number of patients with SCD and CNS crisis was 146 patients, all of them diagnosed by brain magnetic resonance imaging.

Inclusion criteria:

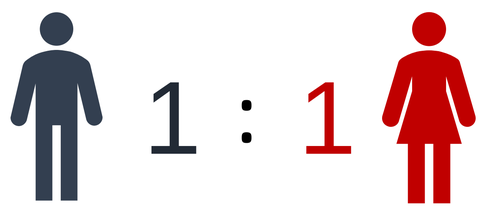
SCD patients with age ≥14 years of any sex presented with any type of CNS crisis.

Exclusion criteria:

SCD patients with age <14 years of any sex and SCD patients **without** evidence of CNS crisis.

RESULTS

146 patient Were included in the study with SCD and CNS crisis



72(49.3%) **MALE**

74(50.7%) **FEMALE**

The age ranges from 14 to 47 years. There is no significant statistical difference between the mean among male versus female SCD patients with CNS crisis. Regarding age groups of patients, higher percentage of them was in the age between 21 and 30 years followed by the age group from 14 to 20 years, without significant difference between both sexes. Most of them were single (76.7%) and non-employed (80.8%) receiving monthly support from government. [Table 1]

Hemoglobin electrophoresis showed (73.3%) of the studied patients to be homozygous SCA (HbSS), while (26.7%) were SC/β-thalassaemia (HbS/ßo and HbS/ß+). The majority of them (89.0%) suffered one CNS crisis, (8.2%) developed twice CNS crises and (2.7%) had three times CNS crises. The age at onset of first CNS crisis was variable, ranging from 6 to 44 years. Half of patients developed the first CNS crisis before 10 years of age (52.1%), and (39.7%) between 10 and 19 years of age.

Table 1. Demographic characteristics of the studied patients with SCD and CNS crisis

Variable	Male (n=72)		Female (n=74)		Total (n=146)		p-value
	№	%	№	%	№	%	
Age group (years):							
≤ 20	28	38.9	25	33.8	53	36.3	0.664
21 - 30	34	47.2	33	44.6	67	45.9	
31 - 40	9	12.5	14	18.9	23	15.8	
> 40	1	1.4	2	2.7	3	2.1	
Marital status:							
Single	53	73.6	59	79.7	112	76.7	0.249
Married	19	26.4	15	20.3	34	23.3	
Employment:							
Employed	8	11.1	1	1.4	9	6.2	0.029*
Non-employed	53	73.6	65	87.8	118	80.8	
Student	11	15.3	8	10.8	19	13.0	

CONCLUSION

Central nervous system insults are common morbidity among SCD patients in Jazan at any age in both sexes with significant post crisis motor disability. The first CNS insult was before 10 years of age. However, with the best management and follow-up provided the outcome of these patients improved and the mortality is decreased.

Footnotes:
References: 1-Berntorp E, Shapiro AD. Modern haemophilia care. Lancet. 2012 Apr 14;379(9824):1447-56. 2-Tonbary YA, Elashry R, Zaki Mel S. Descriptive epidemiology of hemophilia and other coagulation disorders in mansoura, egypt: retrospective analysis. Mediterr J Hematol Infect Dis. 2010 Aug 13;2(3):e2010025. 3-Young G. New challenges in hemophilia: long-term outcomes and complications. Hematol Am Soc Hematol Educ Progr. 2012;2012(1):362–8 Available from: http://www.ncbi.nlm.nih.gov/pubmed/23233605. [cited 2019 Apr 24] 4-Café, A., Carvalho, M., Crato, M. et al. Haemophilia A: health and economic burden of a rare disease in Portugal. Orphanet J Rare Dis 14, 211 (2019). https://doi.org/10.1186/s13023-019-1175-5
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