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ORIGINAL RESEARCH

Impact of Sickle Cell Disease on Academic Performance: A Cross Sectional Study

Mortadah Alsalman[1], Sajjad AlHaddad[2], Ibrahim Alibrahim[3], Abdulhakim Ibrahim Alabdullah[3], Mohammed Hussain Almutawa[3], Abdullah Khalid Alhamam[3], Mohammed Abdullah Albaqshi[3]

[1]Department of Medicine, College of Medicine, King Faisal University, Alahsa, Saudi Arabia; [2]Academy of Family Medicine, Ministry of Health, Alahsa, Saudi Arabia; [3]College of Medicine, King Faisal University, Alahsa, Saudi Arabia

Correspondence: Sajjad AlHaddad, Academy of Family Medicine, Ministry of Health, Alahsa, Saudi Arabia, Tel +966544271184, Email Sajjad-smith@hotmail.com

Purpose: Sickle cell disease (SCD) is a multisystemic disease that results in diverse clinical manifestations in the form of acute and chronic complications. This study aims to assess the academic achievements of patients with SCD.

Methods and Materials: A cross-sectional study was a study conducted among adult patients with SCD from the eastern province of Saudi Arabia, where SCD is more widespread than in other regions.

Results: A total of ninety patients with SCD, whose median age was 32.33 ± 11.84, were retrospectively evaluated. Of the total number, 32 (35.5%) did not obtain a secondary higher education certificate, and five (3.3%) were illiterate. Sixty-three (70%) of the patients were female, indicating no significant association between gender and education level (p-value > 0.05). The patients' mean annual hospitalizations and emergency visits totaling 4.7 and 8.43, respectively, were not significantly associated with education (p-value > 0.05). Throughout the patients' lives, mean blood transfusions and ICU admissions occurred 6.29 and 2.75 times, respectively, which were not significantly associated with education level (p-value > 0.05). However, education was inversely proportional to a history of splenectomy and cholecystectomy ($p < 0.05$). Laboratory parameters (hemoglobin level, WBC, platelets, and hemoglobin F) showed no significant associations with education level (p-value > 0.05). Patients with sickle-thalassemia exhibited a trend of being better-educated than those without the diseases, but the difference did not reach statistical significance (p-value > 0.05). A linear regression analysis revealed no significant associations among clinical and laboratory parameters.

Conclusion: SCD is a multisystemic disease that impacts socioeconomic status, learning ability, and academic achievements. More research is needed to identify factors that predict poor performance. Additionally, intelligence quotient (IQ) and cognitive assessment should be incorporated into early screening programs for SCD in order to implement preventive measures and a thorough investigation of underlying causes.

Keywords: sickle cell disease, academic achievements, education

Introduction

Sickle cell disease (SCD) is an inherited autosomal recessive disorder caused by a mutation in the β-globin gene, and it affects millions of people worldwide.[1,2] About 250,000 people with SCD are born each year. Saudi Arabia is one country with a high prevalence of SCD.[3] Among Saudi children and adolescents, 2400 per 1,000,000 were estimated to have SCD, with the disease being more common in the eastern and southwestern regions of the country.[4,5]

Around twenty years ago, the Saudi Ministry of Education adopted new regulations on special education and emphasized that students with all types of disability should receive their education alongside their peers without disabilities.[6] SCD is a multisystemic disease with a diverse spectrum of clinical manifestations in the form of acute and chronic complications along with increased risk of disabilities and reduced lifespan of affected individuals.[7,8] The most frequent consequences of SCD include acute chest syndrome (ACS), strokes, vaso-occlusive crisis (VOC), invasive infections, and chronic pulmonary hypertension.[9] However, pain is the hallmark of SCD and is the number one cause of hospitalization. Additionally, around 50% of ACS cases occur as a result of painful crisis. Those individuals with both ACS and acute painful crisis had a long hospital stay and an increased risk of readmission, respectively.[10,11] Therefore,

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patients with SCD are at a high risk of absenteeism with an average of 20–40 missed school days per year, which is reflected negatively in school performance.[12,13] Additionally, several studies showed that various biological and environmental factors may affect the cognitive function of patients with SCD, which consequently affects their behavioral and emotional functioning and academic performance.[2,14] Locally, previous study done in the southern region of Saudi Arabia where Benin haplotype is more common showed that SCD has a negative impact on students' academic performance which could be directly or indirectly related to disease complication.[15,16]

To our knowledge, there are a limited number of studies that address the clinical and laboratory factors that influence the academic achievements of patients with SCD. This is of particular importance in our geographic area, where a different SCD genotype, namely, the Arab Indian haplotype, is common and characterized by persistent elevation of hemoglobin F, a well-known modifier of SCD severity. Additionally, SCD severity might be influenced by the coexistence of a common disease in our area, namely, alpha thalassemia that has been found to improve hematological indices and delay the onset of diagnosis, which could contribute ameliorate the general well-being and possibly the survival of patients.[17] Thus, this study aims to assess the academic achievements of patients with SCD along with possible factors that interfere with their performance.

Materials and Methods

This was a cross-sectional study conducted between February to June 2023 in Saudi Arabia's eastern province, where Sickle Cell Disease (SCD) is more common than in other regions. The study consisted of a convenience sample of 90 adult sickle cell patients (18 years and older) who attended the hematology clinic of a tertiary hospital. The inclusion criteria for the study required a confirmed diagnosis of sickle cell disease (HbSS, HbSC, or HbSβ+-thalassemia). Patients were excluded if they were unable to provide informed consent or were not willing to participate in the study.

Data were collected through direct interviews with participants, which were conducted by trained study team members, and a review of the participants' medical records. The data covered the participants' demographic data including gender, level of education, and age, as well as clinical data including the number of hospitalizations and emergency visits per year, previous ICU admissions, frequency of ICU admissions, history of stroke, number of blood transfusions throughout life, history of splenectomy, cholecystectomy, joint replacement, and whether a patient had been started on hydroxyurea or not. Lastly, laboratory data including white blood cells (WBC), hemoglobin (Hb), platelets, MCV, HbA, HbA2, HbS, HbF, and lactate dehydrogenase (LDH) were collected from the patients' most recent laboratory results.

We considered hospitalization frequent if it occurred more than twice per year during the previous two years. On the other hand, we considered blood transfusion significant if it happened more than five times in a patient's life, despite the literature lacking a clear definition of significant blood transfusion in SCD patients. The education system in Saudi Arabia can be divided into two broad categories: general education and higher education. The general education is expected to be completed by the age of eighteen. Therefore, we considered participants to have poor academic achievements if they did not obtain a secondary certificate, the completion of which is expected by the age of eighteen.[18]

Data were collected and analyzed using the International Business Machines (IBM) Statistical Package for the Social Sciences (SPSS) version 26. The descriptive analysis was done to show the mean, median, and mode values with a standard deviation (SD) for quantitative data. We began the analysis by determining the data's normality. The normally distributed data were analyzed, and mean values with a standard deviation were calculated (SD). Pearson correlation analysis and the chi-square test of independence were used to assess any association among different variables. A p-value less than 0.05 was considered significant.

The study was approved by King Faisal University's ethical committee and complied with the Declaration of Helsinki. Also, participants provided their informed consent after being aware of the study's aim, the benefits of conducting the study, confidentiality, and data anonymity.

Results

A total of ninety patients with SCD, whose median age was 32.33 ± 11.84, were evaluated retrospectively. The demographic and general characteristics of the patients are shown in Table 1.

Of the 90 participants, 32 (35.5%) did not obtain a secondary higher education certificate, and five (3.3%) were illiterate. Among all the patients, 63 (70%) were female; however, there was no association between gender and education level (p-value > 0.05). The patients' mean annual hospitalizations and emergency visits totaled 4.7 and 8.43, respectively, although the numbers were not significantly associated with education level (p-value > 0.05). Similarly, the mean number of blood transfusions and intensive care unit admissions throughout the patients' lives amounted to 6.29 and 2.75, respectively; nevertheless, association

Table 1 Patients' Demographic Information and Characteristics (n = 90)

Characteristics	No.	%
Age (Mean ± SD)	32.33 ± 11.84	
Gender		
Male	27	30
Female	63	70
Educational level		
Illiterate	5	5.5
Elementary school	11	12.2
Intermediate school	16	17.8
Secondary higher education	38	42.2
Graduation	17	18.9
Master's degree	3	3.3
Previous ICU admissions	67	74.4
History of stroke	1	1.1
Splenectomy	13	14.4
Cholecystectomy	33	36.7
Joint replacement	5	5.6
On hydroxyurea	42	46.7
Number of blood transfusions throughout life (Mean ± SD)	6.29 ± 4.3	
Frequency of ICU admissions (Mean ± SD)	2.75 ± 3.01	
Number of hospitalizations per year (Mean ± SD)	4.7 ± 4.57	
Number of emergency visits per year (Mean ± SD)	8.43 ± 5.61	
White blood count	10.42 ± 5.1	
Hemoglobin	8.67 ± 1.42	
Platelets	328.84 ± 185.983	
Mean corpuscular volume	78.98 ± 11.62	
Hemoglobin A	8.96 ± 16.64	
Hemoglobin A2	3.29 ± 0.9991	
Hemoglobin S	74.22 ± 14.53	
Hemoglobin F	13.89 ± 6.93	
Lactate dehydrogenase (LDH)	388.94 ± 146.23	

Abbreviations: MCV, mean corpuscular volume; LDH, lactate dehydrogenase.

Table 2 Clinical Data of the Participants (n = 90)

Characteristics	Educational Level						p-value
	Illiterate	Primary	Intermediate	Secondary Higher Education	Graduation	Master	
Gender							
Male	0	2	7	14	2	2	0.077
Female	5	9	9	24	15	1	
Previous ICU admissions							
Yes	4	10	12	27	12	2	0.83
No	1	1	4	11	5	1	
History of stroke							
Yes	0	0	0	0	1	0	0.578
No	5	11	16	38	16	3	
Splenectomy							
Yes	1	4	1	2	4	1	0.035
No	4	7	15	36	13	2	
Cholecystectomy							
Yes	4	3	6	9	10	1	0.047
No	1	8	10	29	7	2	
Joint replacement							
Yes	1	2	0	1	1	0	0.146
No	4	9	16	37	16	3	
Have you been started on hydroxyurea?							
Yes	2	7	7	19	5	2	0.55
No	3	4	9	19	12	1	

Abbreviation: ICU, Intensive Care Unit.

with education level was non-significant (p-value > 0.05). There was no association between education level and hydroxyurea usage, previous surgical intervention, or stroke, but education was inversely proportional to history of splenectomy and cholecystectomy (p-value < 0.05, Table 2). Regarding laboratory parameters, there were no significant associations between education level and hemoglobin level, WBC, platelets, and hemoglobin F (p-value > 0.05). Patients with high hemoglobin A2 or sickle-thalassemia demonstrated better education than others, but the correlation did not reach statistical significance (p-value > 0.05). A linear regression analysis was conducted, but none of the clinical and laboratory parameters were significant.

Discussion

Education is a process and consists of products. Its beneficial effects are pervasive, cumulative, and self-amplifying, growing across the course of life. It is a mixture of knowledge, skills, and capacities that are acquired through experimental and formal learning.[19] Children with chronic illnesses encounter difficulties as they progress in school, with an increased risk of academic underperformance.[20] In our study, we found that more than one-third of participants did not obtain a secondary higher education certificate at an appropriate age, consistent with the performance of patients with SCD and other chronic illnesses.[12,20] Patients with SCD are at a high risk of absenteeism secondary to frequent painful crisis and hospitalization for other acute complications. It has been found that young people with sickle cell disease miss an average of twenty to forty school days in a year.[13] However, frequent emergency visits and hospitalizations were not good predictors of academic performance.

Hemolysis is a fundamental feature of SCD that contributes to the pathophysiology and phenotypic variability of the disease. Both acute and chronic blood transfusions have a critical role in the management and prevention of acute and chronic complications.[21,22] Nevertheless, neither hemoglobin levels nor frequent blood transfusions were correlated with academic performance. Similarly, hydroxyurea plays an essential role in the complex pathophysiology alteration of SCD, resulting in a reduction of acute and long-term complications.[23] However, academic performance was not better among patients who used hydroxyurea compared to those who did not. Additionally, increased fetal hemoglobin is a major genetic modifier of SCD that results in the amelioration of its clinical complications, although its impact on academic performance was not documented either.[24]

This study reveals that the majority of patients with SCD experience poor academic achievement irrespective of absenteeism, which is represented by the number of hospitalizations and emergency visits. Similarly, the limited number of patients with previous strokes suggested that other plausible factors may explain poor academic achievement. Of note, silent strokes are not well documented in our study and could be the hidden cause of academic underperformance. Locally, the prevalence of stroke among individual with SCD range between 6% and 16% and this variation is attributed to the presence of different haplotypes in different area. Unfortunately, the prevalence of silent stroke locally is scarce.[25,26]

Additionally, there were no clear clinical or laboratory predictors of poor academic achievement. However, there are certain limitations that might affect the results of our study. We have no information about the timing of hydroxyurea initiation and whether participants reached the maximum tolerated dose to achieve optimal benefits. Furthermore, school policies twenty year ago might be different from the current ones which might result into variable academic performance between previous and current generations affected with SCD. Lastly, other socioeconomic factors and chronic pain influences and impacts were not fully addressed in the study. Further studies are needed to study the impact of early intervention via different therapeutic options including hydroxyurea and stem cell transplantation on the amelioration of academic performance and achievements.

Conclusion

Sickle cell disease is a multisystemic disease, but its impacts extend to the affected patients' socioeconomic status—particularly learning ability and academic achievements. However, the lack of obvious clinical or laboratory predictors of poor academic performance could result into suboptimal intervention. Therefore, further studies are needed to explore factors that predict poor performance. Additionally, silent stroke surveillance, intelligence quotient (IQ) and cognitive assessment programs should be incorporated into early screening programs for patients with SCD in order to implement early preventive measures and a thorough work-up for underlying perceivable causes. Furthermore, new school policies should be established to accommodate individuals with frequent medical absences.

Abbreviations

SCD, sickle cell disease; WBC, white blood cell; IQ, intelligence quotient; ACS, acute chest syndrome; VOC, vaso-occlusive crisis; Hb, hemoglobin; LDH, lactate dehydrogenase; IBM, International Business Machine; SPSS, Statistical Package for the Social Sciences; SD, standard deviation.

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Disclosure

The authors declare no conflicts of interest in this work.

References

1. Colombatti R, Casale M, Russo G. Disease burden and quality of life of in children with sickle cell disease in Italy: time to be considered a priority. *Ital J Pediatr*. 2021;47(1). doi:10.1186/s13052-021-01109-1

2. Prussien KV, Siciliano RE, Ciriegio AE, et al. Correlates of cognitive function in sickle cell disease: a meta-analysis. *J Pediatr Psychol*. 2020;45(2):145–155. doi:10.1093/jpepsy/jsz100
3. Alsaeed ES, Farhat GN, Assiri AM, et al. Distribution of hemoglobinopathy disorders in Saudi Arabia based on data from the premarital screening and genetic counseling program, 2011–2015. *J Epidemiol Glob Health*. 2018;7(S1):S41–7. doi:10.1016/j.jegh.2017.12.001
4. AlHamdan NAR, AlMazrou YY, AlSwaidi FM, Choudhry AJ. Premarital screening for thalassemia and sickle cell disease in Saudi Arabia. *Genet Med*. 2007;9(6):372–377. doi:10.1097/GIM.0b013e318065a9e8
5. Zuair A, Bin AS, Alhumaidi R, Alrabiah M, Alshabanat A. The burden of sickle cell disease in Saudi Arabia: a single-institution large retrospective study. *Int J Gen Med*. 2023;16:161–171. doi:10.2147/IJGM.S393233
6. Almalky HA, Alwahbi AA. Teachers' perceptions of their experience with inclusive education practices in Saudi Arabia. *Res Dev Disabil*. 2023;140:104584. doi:10.1016/j.ridd.2023.104584
7. McGann PT, Ware RE. Hydroxyurea for sickle cell anemia: what have we learned and what questions still remain? *Curr Opin Hematol*. 2011;18(3):158–165. doi:10.1097/MOH.0b013e32834521dd
8. Carden MA, Little J. Emerging disease-modifying therapies for sickle cell disease. In: *Haematologica*. Vol. 104. Ferrata Storti Foundation; 2019:1710–1719.
9. Shah N, Bhor M, Xie L, Paulose J, Yuce H, Kamolz L-P. Sickle cell disease complications: prevalence and resource utilization. *PLoS One*. 2019;14(7):e0214355. doi:10.1371/journal.pone.0214355
10. El-Ghany SM A, Tabbakh AT, Nur KI, Abdelrahman RY, Etarji SM, Almuzaini BY. Analysis of causes of hospitalization among children with sickle cell disease in a group of private hospitals in jeddah, Saudi Arabia. *J Blood Med*. 2021;12:733–740. doi:10.2147/JBM.S318824
11. Ballas SK, Gupta K, Adams-Graves P. Sickle cell pain: a critical reappraisal. *Blood*. 2012;120(18):3647–3656. doi:10.1182/blood-2012-04-383430
12. Al-Saqladi AW. The impact of sickle cell disease severity on school performance in affected Yemeni children. *J Appl Hematol*. 2016;7(4):124. doi:10.4103/1658-5127.198506
13. Schwartz LA, Radcliffe J, Barakat LP. Associates of school absenteeism in adolescents with sickle cell disease. *Pediatr Blood Cancer*. 2009;52(1):92–96. doi:10.1002/pbc.21819
14. Berkelhammer LD, Williamson AL, Sanford SD, et al. Neurocognitive sequelae of pediatric sickle cell disease: a review of the literature. *Child Neuropsychol*. 2007;13(2):120–131. doi:10.1080/09297040600800956
15. Jastaniah W. Epidemiology of sickle cell disease in Saudi Arabia. *Ann Saudi Med*. 2011;31(3):289–293. doi:10.4103/0256-4947.81540
16. Alhazmi A, Hakami K, Abusageah F, et al. The impact of sickle cell disease on academic performance among affected students. *Children*. 2022;9(1):15.
17. Rumaney MB, Ngo Bitoungui VJ, Vorster AA, et al. The co-inheritance of alpha-thalassemia and sickle cell anemia is associated with better hematological indices and lower consultations rate in Cameroonian patients and could improve their survival. *PLoS One*. 2014;9(6):e100516. doi:10.1371/journal.pone.0100516
18. Rabaah A, Doaa D, Asma A. Early childhood education in Saudi Arabia: report. *World J Educ*. 2016;6(5). doi:10.5430/wje.v6n5p1
19. Hahn RA, Truman BI. Education improves public health and promotes health equity. *Int J Health Serv*. 2015;45(4):657–678. doi:10.1177/0020731415585986
20. Hu N, Fardell J, Wakefield CE, et al. School academic performance of children hospitalised with a chronic condition. *Arch Dis Child*. 2022;107(3):289–296. doi:10.1136/archdischild-2020-321285
21. Kato GJ, Steinberg MH, Gladwin MT. Intravascular hemolysis and the pathophysiology of sickle cell disease. *J Clin Invest*. 2017;127(3):750–760. doi:10.1172/JCI89741
22. Davis BA, Allard S, Qureshi A, et al. Guidelines on red cell transfusion in sickle cell disease Part II: indications for transfusion. *Br J Haematol*. 2017;176(2):192–209. doi:10.1111/bjh.14383
23. Alsalman M, Alkhalifa H, Alkhalifa A, et al. Hydroxyurea usage awareness among patients with sickle-cell disease in Saudi Arabia. *Health Sci Rep*. 2021;4(4). doi:10.1002/hsr2.437
24. Paikari A, Sheehan VA. Fetal haemoglobin induction in sickle cell disease. In: *British Journal of Haematology*. Vol. 180. Blackwell Publishing Ltd; 2018:189–200.
25. Babeer G, Omran D, Bawahab N, et al. Prevalence and risk factors of stroke among children with sickle cell disease: a retrospective study at a tertiary care center. *Cureus*. 2023. doi:10.7759/cureus.41960
26. Alsultan A, Alabdulaali MK, Griffin PJ, et al. Sickle cell disease in Saudi Arabia: the phenotype in adults with the Arab-Indian haplotype is not benign. *Br J Haematol*. 2014;164(4):597–604. doi:10.1111/bjh.12650