School performance of children with sickle cell disease in Basra, Iraq

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Abstract:
BACKGROUND: Children with sickle cell disease (SCD) have a greater likelihood of demonstrating many medical complications that may put them at risk for a variety of difficulties, including poor school performance and cognitive impairment.

OBJECTIVES: This study was designed to assess the school performance of primary school-aged patients with SCD compared to age- and gender-matched healthy students and to evaluate the factors that influence school performance in these patients.

MATERIALS AND METHODS: A total of 68 patients with SCD from 48 primary schools and 68 healthy student classmates (control group), aged 7–12 years, were recruited. The average school examination scores were used as a school performance measure, while cognitive functioning was assessed by using the Draw-A-Person test.

RESULTS: The average school examination scores and cognitive function scores of patients with SCD (82.46 ± 15.48 and 24.72 ± 7.48, respectively) were significantly lower than the corresponding scores of their classmate control group (93.42 ± 10.02 and 27.84 ± 7.46, respectively), P < 0.05. Students with SCD missed significantly more school days (12.37 ± 10.57) than healthy students (3.18 ± 3.62), P < 0.001, and high school absence was significantly associated with low school performance among SCD patients, P = 0.027. Among patients with SCD, Pearson correlation revealed a negative association between school performance and age and school absence days (r = −0.346 and r = −0.390, respectively, P < 0.01) and a positive association between school performance and maternal education (r = −0.388, P < 0.001).

CONCLUSIONS: School performance and cognitive function were significantly lower among patients with SCD than among their classmates, and school absence and the age of patients had a negative impact on school performance.

Keywords: Children, cognitive function, school absence, school performance, sickle cell disease

Introduction

Education is one of the most important aspects of human development. Every child should have the opportunity to achieve his or her academic potential. Poor school performance should be seen as a “symptom” reflecting a larger underlying problem in children, and it can cause significant stress for parents.[1]

Patients with sickle cell disease (SCD) have been identified as a chronic illness population with a greater likelihood of demonstrating many medical complications that may put the child at risk for a variety of difficulties, including poor school performance and cognitive impairment sequelae.[2]

Many factors predict school performance in the SCD population. Understanding the factors that influence school performance in children with SCD will allow professionals to best target their efforts toward serving these children and addressing their educational needs.[3,4]

These factors may include the cognitive ability of SCD patients, as different studies...
have discussed the relationships between the cognitive ability of SCD patients and their school performance and revealed poor school performance among children with SCD. However, other studies have not found any differences between SCD patients and healthy students in the areas of cognitive function and school performance.

The mechanisms that underlie cognitive impairment in patients with SCD are poorly understood. Although neurological accidents (stroke, silent infarcts, and transient ischemic attacks) are among the most worrisome clinical complications of SCD and are frequently accompanied by cognitive impairment, it is not known whether cognitive deficits in patients with SCD occur only due to stroke or whether impairment can also occur among children who have no evidence of focal brain injury.

The cognitive impairments in children with SCD are characterized by a reduction in general intellectual functioning, language and verbal abilities, visual-motor and visual-locational processing, memory, and school performance. School absence is another factor; patients with SCD have been found to miss an average of 20–40 more school days per year than healthy students, which can deteriorate school examination scores and lead to poorer school performance in comparison with healthy students. School absence seems to be due mainly to the medical complications associated with SCD, which lead to repeated hospitalizations and frequent medical consultations.

Poor school performance has been documented to be high among children from low socioeconomic status (SES) backgrounds. Poor school performance may be the result of the interplay between the child and environmental factors. Patients with unsuitable home environments, little social support, and families of low income are more likely to exhibit poorer performance on cognitive testing, to be retained a grade level, or to not complete high school. Furthermore, reduced support from parents with lower educational levels is associated with repeating a grade and poor school performance and vice versa. It is also believed that learners benefit from the presence of parents who are educated themselves.

Although no significant gender differences in general cognitive abilities have been found, it has been reported that boys may experience grade failure more than girls among children with SCD. Girls with SCD have been found to be more successful in school than boys with the same disease. Compared to healthy peers, children with SCD have been rated by their caregivers to have lower physical, psychological, and social health-related quality of life, with associated limitations in self-esteem, school and social participation, and general psychological adjustment, which lead to poor school performance.

Based on the reviewed literature, we hypothesized that school performance and cognitive functioning would be significantly impaired among children with SCD. We further hypothesized that school absenteeism, female gender, and low SES would be associated with poor academic functioning in children with SCD.

This study is one of the first studies in Iraq to describe the school performance of children with SCD and to examine the potential factors contributing to academic achievement in these children.

**Materials and Methods**

This case–control study was conducted in patients with SCD who had been registered at the Center for Hereditary Blood Diseases (CHBD) over the period from November 2015 to January 2016. It included children aged 7–12 years. A total of 68 patients (34 females and 34 males) from 48 primary schools (39 public and 9 private) were recruited, along with 68 age- and gender-matched classmates from the same primary schools who were free of any chronic conditions (control group). The controls sat next to the subjects in class.

In addition to age and gender, further information that was collected included the educational levels of the parents, the type of school (public or private), the presence of a repeated grade, and SES. SES was scored according to the presence of an owned or rented house (scored as 0 or 1), the presence or absence of a private car (scored as 0 or 1), and the number of electric appliances (as air conditioner, refrigerator, television, and electric water boiler) in the household (up to 2 = 1, 3–5 = 2, 6 or more = 3). An overall SES score was calculated for each subject; a score of 2 or less indicated low SES, a score of 3–4 indicated middle SES, and a score of >4 was considered high SES.

For both cases and controls, the following information was recorded: The number of absent school days and the average of school examination scores documented from their school’s record of the previous year, with the date of a school visit.

The frequencies of hospital admission, causes of hospitalization, frequency of vaso-occlusive crises (VOC), frequency of blood transfusion (BT), history of stroke, acute chest syndrome (ACS), and
avascular bone necrosis (AVN) during the academic year of study were also recorded for patients from the files at the CHBD.

The severity of SCD was assessed according to the frequency of VOC requiring hospitalization per year, frequency of BT per year, history of stroke, ACS, and/or AVN. The patient was classified to have severe disease when they had a high frequency of VOC requiring hospitalization (≥3/year), a high frequency of BT (≥3/year), at least one stroke, ACS, and/or AVN.\(^{[23,24]}\)

The type of SCD also reflected the severity of disease, in which those with SCA and Hb-S/\(\beta^0\) thalassemia were regarded to have severe disease, while those with other types, such as Hb-S/\(\beta^+\) thalassemia, had milder disease.\(^{[25,26]}\)

Patient data were collected by direct interview with the patients and one of their parents, who had consulted the CHBD for routine follow-up. For the control group, data were collected by visiting the same 48 primary schools of patients with SCD.

Ethical permission to conduct the research was obtained from the CHBD and all participants before the study began.

**School performance measure**
The participants’ school performance, which was obtained through calculating the average of three examination results (first semester, second semester, and final examination) for each educational subject from the school’s documentation of the previous academic year (2014/2015), was used for both cases and controls.\(^{[11,27]}\)

This average was further graded as high (≥75%), moderate (50% to 75%), and low (<50%).\(^{[11]}\)

**School absence**
The total number of days of school absence for the 2014/2015 academic year was obtained for each pair of students from the class attendance registry. The number of days was further classified into high school absence (>12) and low school absence (≤12).\(^{[11,28]}\)

**Cognitive function**
Although there are several different standardized tests designed to assess relative intelligence as a measure of general cognitive ability (e.g., Stanford-Binet Intelligence Scales and Wechsler Intelligence Scale for Children). However, these were not available. Therefore, cognitive function was assessed using the Draw-a-Person test (DAPT) scoring system, which was proposed by Ziler.\(^{[27,28]}\)

However, the assessment of cognitive functioning using a global measure, such as the DAPT test, is not sufficient because children with SCD may suffer from specific cognitive impairments; nonetheless, this measure was chosen because it is more accessible in Iraq than other assessment instruments.

**Statistical analysis**
Statistical analysis was done using SPSS program version 20 software, (IBM, Armonk, NY, USA). The data were expressed as the means ± standard deviations. Comparisons of proportions were performed by crosstabs using Chi-square tests and Fisher’s exact tests. The \(t\)-test was used for the quantitative comparison between two means of different samples. Comparisons between groups were made using the one-way analysis of variance (ANOVA). \(P < 0.05\) was considered statistically significant for the above tests.

Pearson correlation was used to determine the association between school performance and different variables among patients with SCD. Given that significant differences in SES and maternal education between SCD patients and healthy controls were observed and that age, school absence days, and DAPT scores were related to school performance, we included these factors as covariates on school performance.

**Results**
A total of 68 patients with SCD and 68 age- and sex-matched healthy students were included in this study. Their ages ranged from 7 to 12 years, with a mean age of 9.37 ± 1.39 years for patients with SCD and 9.35 ± 1.38 years for the control group.

Subjects were subdivided into three subgroups by age: 7–8 years, 9–10 years, and 11–12 years for both the case and control groups. The sex ratio was equal for both cases and controls.

Fifty-nine subjects were recruited from public schools and 9 from private schools.

The study demonstrated that a significantly higher number of children within the control group belonged to middle- and high-SES families, while higher numbers of patients with SCD were from families of low- and middle-SES backgrounds, \(P < 0.05\).

In addition, the maternal educational level of healthy children was found to be significantly higher than that of the patient group, \(P < 0.05\), [Table 1].

The study also demonstrated that both the average school examination scores and DAPT scores were significantly
lower in patients with SCD compared to the control group \( (P < 0.05) \). Furthermore, students with SCD missed significantly more school days than healthy students, \( P < 0.05 \), [Table 1].

The study found that both males and females with SCD showed significantly lower school performance compared to control group members of the same gender \( (P < 0.05) \) [Figure 1a]. In addition, SCD patients who were 11–12 years old demonstrated a significantly lower average school examination score than the other two age groups, \( P < 0.05 \). However, healthy students who were 9–10 years old and 11–12 years old showed significantly higher scores than their SCD peers \( (P < 0.05) \) [Figure 1b].

Concerning the DAPT scores, females showed significantly higher scores than males in both the patient and control groups \( (P < 0.05) \), while among males, healthy children showed significantly higher scores than SCD patients \( (P < 0.05) \) [Figure 1b].

The study did not report a significant difference in DAPT scores among different age groups between the patient and control groups and among the age groups of SCD patients \( (P > 0.05) \) [Figure 2b].

The mean number of school absence days for both male and female SCD patients were significantly higher than those for the control group \( (P < 0.05) \) [Figure 3a].

The mean number of school absence days for SCD patients who were 11–12 years old was significantly higher than for the other two age groups, \( P < 0.05 \). Patients with SCD, those who were 9–10 years old and 11–12 years old showed a significantly higher number of school absence days than their control peers \( (P < 0.05) \) [Figure 3b].

School performance grades in relation to selected sociodemographic and clinical variables were studied. While none of the sociodemographic variables showed

| Variable                                           | SCD patients | Control group | \( P \) value |
|----------------------------------------------------|--------------|---------------|--------------|
| Socioeconomic status, n (%)                        |              |               |              |
| Low                                                | 13 (19.12)   | 8 (11.76)     | 0.010*       |
| Middle                                             | 46 (67.64)   | 36 (52.94)    |              |
| High                                               | 9 (13.24)    | 24 (35.30)    |              |
| Educational level of father, n (%)                 |              |               |              |
| Primary                                            | 21 (30.88)   | 27 (39.71)    | 0.369*       |
| Intermediate                                       | 20 (29.41)   | 13 (19.12)    |              |
| Secondary                                          | 11 (16.18)   | 8 (11.76)     |              |
| Higher education                                   | 16 (23.53)   | 20 (29.41)    |              |
| Educational level of mother, n (%)                 |              |               |              |
| Primary                                            | 30 (44.11)   | 22 (32.35)    | 0.022*       |
| Intermediate                                       | 17 (25.00)   | 19 (27.95)    |              |
| Secondary                                          | 12 (17.65)   | 5 (7.35)      |              |
| Higher education                                   | 9 (13.24)    | 22 (32.35)    |              |
| Average school examination scores (mean±SD)        | 82.46±15.48  | 93.42±10.02   | <0.001**     |
| DAPT scores (mean±SD)                              | 24.72±7.48   | 27.84±7.46    | 0.016**      |
| School absence days (mean±SD)                      | 12.37±10.57  | 3.18±3.62     | <0.001**     |

*A Chi-square test is used to assess the \( P \) value, **An independent \( t \)-test is used to assess the \( P \) value. DAPT=Draw-a-Person test, SD=Standard deviation, SCD=Sickle cell disease

**Table 1: Selected sociodemographic variables, average school examination scores, Draw-a-Person test scores, and school absence days of the studied population**
significant differences in relation to school performance grades ($P > 0.05$), patients who were in the 9- to 10-year-old age group had higher school performance grades than the other two age groups, $P < 0.05$. In addition, SCD patients activated with a low number of school absence days had higher school performance grades compared to those with a high number of school absence days, $P < 0.05$, [Table 2]. The selected variables for clinical severity measures (number of VOC/year, BT/year, and ACS) showed no impact on the school performance grades of patients with SCD, $P > 0.05$, [Table 3].

Pearson correlation coefficient revealed that the school performance of patients with SCD was negatively associated with the number of school absence days and the age of patients, $P < 0.05$, while the educational level of mothers of patients with SCD was positively associated with the school performance of their children, $P < 0.01$, while both SES and DAPT scores were not found to be associated with school performance of SCD patients [Table 4].

**Discussion**

In the current study, the mean of the school examination scores of patients with SCD were lower than that of the control group comprised of their classmates, which is consistent with the findings of Schatz,$^{[29]}$ who found that children with SCD had a higher rate of academic achievement problems and more frequent instances of multiple grade repetitions compared to controls. However, in contrast to our findings, Ezenwosu et al. in Nigeria found no significant differences in the mean overall academic scores of subjects and controls.$^{[11]}$ This discrepancy in overall academic scores may be related to different rates of school absences among the studied patients.

Impaired cognitive functioning is one of the most significant developmental outcomes faced by individuals with SCD. Furthermore, there has been growing evidence that children with SCD experience cognitive deficits across several domains when compared to healthy peers.$^{[30]}$

In the current study, the cognitive function scores, measured by DAPT scores, of patients with SCD were lower than the scores of the control group. Similar to this finding, Noll et al. found that children with SCD have lower cognitive functioning than the control group.$^{[6]}$ In addition, Wang et al. in the USA found that school-aged children with SCD had compromised cognition in certain areas of functioning over time.$^{[8]}$ Although it is well established that children with SCD are at risk for significant decrements in cognitive functioning, there is limited understanding of the underlying causes.$^{[30]}$
Table 2: School performance grades of patients with SCD in relation to selected socio-demographic variables

| Variables                      | School performance grades | Total | P-value |
|--------------------------------|---------------------------|-------|---------|
|                                | Low (2.94)                |       |         |
|                                | Moderate (26.47)          |       |         |
|                                | High (70.59)              |       |         |
| Age (years)                    |                           |       |         |
| 7-8                            | 0 (0.00)                  | 4 (5.88) | 17 (25.00) | 21 (30.88) | 0.017 |
| 9-10                           | 1 (1.47)                  | 6 (8.82) | 26 (38.24) | 33 (48.53) |       |
| 11-12                          | 1 (1.47)                  | 8 (11.76) | 5 (7.35)   | 14 (20.59) |       |
| Gender                         |                           |       |         |
| Male                           | 1 (1.47)                  | 10 (14.71) | 23 (33.82) | 34 (50.00) | 0.890*|
| Female                         | 1 (1.47)                  | 8 (11.76) | 25 (36.76) | 34 (50.00) |       |
| Socio-economic status          |                           |       |         |
| Low                            | 0 (0.00)                  | 5 (7.35)   | 8 (11.76)  | 13 (19.12) | 0.138 |
| Middle                         | 1 (1.47)                  | 13 (19.12) | 32 (47.06) | 46 (67.64) |       |
| High                           | 1 (1.47)                  | 0 (0.00)   | 8 (11.76)  | 9 (13.24)  |       |
| Educational level of father    |                           |       |         |
| Primary                        | 1 (1.47)                  | 6 (8.82)   | 14 (20.59) | 21 (30.88) | 0.691 |
| Intermediary                   | 1 (1.47)                  | 6 (8.82)   | 13 (19.12) | 20 (29.41) |       |
| Secondary                      | 0 (0.00)                  | 4 (5.88)   | 7 (10.29)  | 11 (16.18) |       |
| Higher education               | 0 (0.00)                  | 2 (2.94)   | 14 (20.59) | 16 (23.53) |       |
| Educational level of mother    |                           |       |         |
| Primary                        | 1 (1.47)                  | 8 (11.76)  | 21 (30.88) | 30 (44.12) | 0.505 |
| Intermediary                   | 1 (1.47)                  | 7 (10.29)  | 9 (13.24)  | 17 (25.00) |       |
| Secondary                      | 0 (0.00)                  | 2 (2.94)   | 10 (14.71) | 12 (17.65) |       |
| Higher education               | 0 (0.00)                  | 1 (1.47)   | 8 (11.76)  | 9 (13.24)  |       |
| School absence days            |                           |       |         |
| Low                            | 0 (0.00)                  | 8 (11.76)  | 33 (48.53) | 41 (60.29) | 0.027 |
| High                           | 2 (2.94)                  | 10 (14.71) | 15 (22.06) | 27 (39.71) |       |
| Type of school                 |                           |       |         |
| Public                         | 2 (2.94)                  | 16 (23.53) | 41 (60.29) | 59 (86.76) | 1.000 |
| Private                        | 0 (0.00)                  | 2 (2.94)   | 7 (10.29)  | 9 (13.24)  |       |

Fishers exact test was used to assess P value

Table 3: School performance grades of patients with SCD in relation to selected clinical variables

| Variables      | School performance grades | Total | P-value |
|----------------|---------------------------|-------|---------|
|                | Low (2.94)                |       |         |
|                | Moderate (26.47)          |       |         |
|                | High (70.59)              |       |         |
| VOC/Year       |                           |       |         |
| 0              | 1 (1.47)                  | 17 (25)   | 34 (50)  | 52 (75.47) | 0.293 |
| 1 - 2          | 1 (1.47)                  | 1 (1.47)  | 13 (19.12) | 15 (22.06) |       |
| ≥ 3            | 0 (0.00)                  | 0 (0.00)  | 1 (1.47)  | 1 (1.47)   |       |
| Blood transfusion/year |                   |       |         |
| 0              | 1 (1.47)                  | 16 (23.53) | 37 (54.42) | 54 (79.42) | 0.426 |
| 1 - 2          | 0 (0.00)                  | 1 (1.47)  | 6 (8.82)  | 7 (10.29)  |       |
| ≥ 3            | 1 (1.47)                  | 1 (1.47)  | 5 (7.35)  | 7 (10.29)  |       |
| ACS            |                           |       |         |
| Yes            | 0 (0.00)                  | 0 (0.00)  | 4 (5.88)  | 4 (5.88)   | 0.618 |
| No             | 2 (2.94)                  | 18 (26.47) | 44 (64.71) | 64 (94.12) |       |

P-value is assessed by Fisher's exact test. VOC; vaso-occlusive crises, ACS; acute chest syndrome

cognitive decrement may be related to the direct effects of SCD on brain functioning, such as cerebral infarction, or indirect effects related to social or environmental disadvantages (e.g., decreased learning opportunities and increased physical limitations from chronic illness). The cognitive function measured by DAPT scores showed that females have a significantly higher score than males in both the patient and control groups. The reasons for these gender differences in cognitive function have not yet been clarified, but it has been indicated that specific components of self-regulation, behavioral
regulation, or self-regulated learning could contribute to gender differences in cognitive function.\cite{20}

The present study showed that students with SCD have a higher number of school absence days than healthy students, consistent with findings from Ogunfowora et al.\cite{10} in Nigeria, who found that patients with SCD have more school absence days than healthy peers and findings from Schwartz et al.\cite{13} in the USA, who found a significant problem of school absenteeism among patients with SCD. The high absence rate in children with SCD may be due to many factors. These factors include frequent routine follow-up visits, psycho-emotional disturbances, and recurrent crises resulting in frequent hospitalization.\cite{11}

School absenteeism may be a major determinant contributing to decreased school performance in SCD children.

The present study reported that patients with SCD who had less frequent school absence days showed significantly higher school performance grades compared to those with more frequent school absence days. Similar to our findings, Crosby et al. found a negative impact of absenteeism on the school performance grades of SCD patients, although the participants were adolescents aged 12–20 years.\cite{3} During these absences, students are missing vital academic content. Difficulties associated with missed instruction are further complicated by subject matter that builds on previous knowledge, such as mathematics. In addition, children may miss exams and standardized tests.\cite{1}.

Severity measures in SCD may be related to clinical factors. In this study, none of the clinical severity measures had an impact on school performance grades in children because only a minority of patients met the clinical criteria that reflected disease severity. A previous study in Basra revealed that the mean fetal hemoglobin level was 19.65 ± 7.42, with significant negative association reported between Hb F level and frequency of painful crisis, ACS and frequency of hospitalizations.\cite{32} In contrast to the findings of the current study Crosby et al. reported that patients with a severe form of SCD may miss school due to pain episodes or organ and/or bone damage.\cite{3}

An earlier study on SCD patients in Basra found that most of the caregivers (67.7\%) of children with SCD lost income due to time-consuming caring for their children with SCD which significantly contributes to the financial impact practiced by caregivers and adversely impact the SES of such families.\cite{33}

In terms of sociodemographic variables, although a significantly higher number of children with SCD patients came from lower SES backgrounds than the control group, this study documented no effects of SES of family on school performance. However, King et al.\cite{14} in the USA and Ezenwosu et al.\cite{11} in Nigeria documented a significant association between academic score ratings and SES and attributed this association to poor motivation, unsatisfactory home environment.\cite{11} The finding of our study concerning SES may be attributed to the fact that the majority of participants in this study attended public schools, in which the education is free of charge for pupils; in addition, in Iraq, health services provided are free of cost for all patients. In addition, the SES classification utilized in the current study did not include parental education for the SES scoring, as in many other studies and this probably led to a discrepancy in the results concerning the impact of SES on school performance.

Maternal education levels were found to be positively correlated with school performance in children with SCD. This finding is consistent with those of Smith et al.\cite{4} who found that parent education is positively correlated with school performance in children with SCD, mainly because less educated parents may have fewer opportunities for learning and stimulation. In addition, in our society, this finding can be explained by the idea that fathers take care of the living expenses and have less time than mothers to monitor the duties of their children.

The current study showed a decline of school performance with increasing age in children with SCD, which is consistent with the findings of Wang et al.\cite{8} and Ezenwosu et al.\cite{11}. This finding could be attributed to the greater network of brain regions activated during processing tasks and mental activities exhibited by younger children than older children.\cite{34} Another plausible explanation could be that older children are faced with more problems, including burdensome homework, overscheduled activities, and television viewing, which might cause sleep disturbances and consequent lower cognitive function.\cite{12}
Conclusions

This study suggests that the academic performance of children with SCD and absenteeism are strongly related and that students with SCD are absent from school more often than those without SCD, which may be a major factor contributing to decreased scores on tests. In addition, factors related to social or environmental disadvantages (age of the patient and maternal education) have also been reported as potential factors. Therefore, extra academic programs are required for children with SCD who have poor school performance, and regular evaluation of intelligence in follow-up clinics is also necessary to detect any early deviations from normal development that may require early medical intervention.

Acknowledgments

We would like to thank Dr. Jassim Naeem, Professor at the Department of Community Medicine, College of Medicine, Basra University, and Dr. Zeina Anes Salman (Pediatrician) and Dr. Ammar Ali Hussain (Pediatrician) at Basra Maternity and Children Hospital for their great assistance in carrying out the statistical analysis of data.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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