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Prevalence of Splenic Sequestration in Patient with Sickle Cell Disease in Hereditary Blood Disease Center in Kerbela Provence

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Abstract

Background: Sickle cell disease is a genetic disorder in hemoglobin characterized by chronic hemolytic anemia and vaso-occlusive complications. splenic sequestration is one of the most serious early-life complications, a life-threatening condition resulting from the rapid pooling of sickled red blood cells in the spleen, leading to acute anemia and hypovolemic shock. This condition primarily affects children under 5 years and carries high recurrence and mortality rates.

Methodology: This is retrospective cross-sectional study that conducted 101 patients with sickle cell disease from August 2023 to October 2024. The study took place Hereditary Hematological center in Karbala Provence. Data were collected using a structured questionnaire

Results: A total of 101 sickle cell disease patients with mean age 7 ± 2.37 years were included. Most were aged 7–10 years (63.4%) and male account (56.4%), with 65.3% reporting parental consanguinity. No significant association was found between sociodemographic factors and splenic sequestration. The majority of splenic sequestration cases were diagnosed between ages 1–3 years (78.3%). Although frequent sequestration episodes were more common in older children, the association with age group was not significant. A significant association was observed between splenic sequestration and disease type ($p = 0.027$), with SS-type accounting for 73.9% of cases. Splenectomy was significantly associated with frequent sequestration episodes ($p = 0.005$). Hydroxyurea use showed no significant link with sequestration, while regular blood transfusion was strongly associated ($p = 0.001$).

Conclusion: Splenic sequestration is a significant early complication in children with sickle cell disease, especially those with the SS genotype. Most cases occurred between ages 1–3, with no significant link to sociodemographic factors. Frequent sequestration episodes were strongly associated with the need for splenectomy, while regular blood transfusion was also significantly linked to sequestration. In contrast, hydroxyurea use showed no significant association.

انتشار احتشاء الطحال في المرضى المصابين بمرض الخلايا المنجلية في مركز أمراض الدم الوراثية بمحافظة كربلاء

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الخلاصة

المقدمة: يُعدّ فقر الدم المنجلي اضطرابًا وراثيًا في الهيموغلوبين يتميّز بفقر دم انحلالي مزمن ومضاعفات انسدادية وعائية. ويُعدّ احتجاز الطحال من أخطر المضاعفات المبكرة في مراحل الطفولة، وهو حالة مهدِّدة للحياة تنجم عن التجمّع السريع لكريات الدم الحمراء المنجلية داخل الطحال، مما يؤدي إلى فقر دم حاد وصدمة نقص حجم الدم. تُصيب هذه الحالة غالبًا الأطفال دون سن الخامسة، وترتبط بمعدلات مرتفعة من النكس والوفيات.

العينات وطرق العمل: أُجريت هذه الدراسة المقطعية المستعرضة بأثرٍ رجعي على **101**مريضًا مصابًا بمرض الخلايا المنجلية خلال الفترة من أغسطس 2023 إلى أكتوبر 2024 .نُفِّذت الدراسة في مركز أمراض الدم الوراثية بمحافظة كربلاء .جُمعت البيانات باستخدام استبيان مُنظّم.

النتائج: شملت الدراسة 101مريضًا بمرض الخلايا المنجلية، وكان متوسط العمر 2.37 ± 7سنة. كانت الفئة العمرية الأكثر شيوعًا 10–7سنوات بنسبة(63.4%) ، وبلغت نسبة الذكور (56.4%) ، مع وجود قرابة أبوية (زواج الأقارب) لدى (65.3%) من الحالات. لم تُسجَّل علاقة ذات دلالة إحصائية بين العوامل الاجتماعية والديموغرافية وحدوث احتجاز الطحال. تم تشخيص معظم حالات احتجاز الطحال ضمن الفئة العمرية 3–1سنوات بنسبة .(78.3%) وعلى الرغم من أن تكرار نوبات الاحتجاز كان أعلى لدى الأطفال الأكبر سنًا، فإن العلاقة مع الفئات العمرية لم تكن ذات دلالة إحصائية. وُجدت علاقة ذات دلالة إحصائية بين احتجاز الطحال ونوع المرض($p = 0.027$) ، حيث شكّل النمط الجيني SSنسبة (73.9%) من الحالات. كما ارتبط استئصال الطحال بشكلٍ معنوي بتكرار نوبات الاحتجاز .($p = 0.005$) ولم يُظهر استخدام الهيدروكسي يوريا علاقة ذات دلالة إحصائية مع احتجاز الطحال، في حين ارتبط نقل الدم المنتظم ارتباطًا قويًا وحدوث احتجاز الطحال.($p = 0.001$)

الاستنتاج:

يُعدّ احتجاز الطحال من المضاعفات المبكرة المهمة لدى الأطفال المصابين بمرض الخلايا المنجلية، ولا سيّما لدى المصابين بالنمط الجيني .SSحدثت معظم الحالات بعمر 3–1سنوات دون وجود علاقة معنوية بالعوامل الاجتماعية والديموغرافية. كما ارتبط تكرار نوبات الاحتجاز بقوة بالحاجة إلى استئصال الطحال، في حين كان نقل الدم المنتظم مرتبطًا بشكلٍ معنوي بحدوث احتجاز الطحال. في المقابل، لم يُظهر الهيدروكسي يوريا علاقة ذات دلالة إحصائية مع احتجاز الطحال.

1. Introduction

Sickle cell disease (SCD) is a genetic disorder caused by a single base-pair mutation in the hemoglobin gene, which results in the production of sickle hemoglobin (HbS). This mutation is inherited in an autosomal recessive pattern, either from two HbS alleles or in combination with other hemoglobin variants such as HbC or β-thalassemia (Crossley et al., 2022; Taher et al., 2025). Sickle cell disease is a common inherited disorder and multisystem involvement marked by chronic hemolytic anemia and recurrent vaso-occlusive phenomena driven by endothelial activation and dysregulation of coagulation and fibrinolytic pathways (Lizarralde-Iragorri & Shet, 2020;Ofori-Acquah, 2020; Odame, 2023). splenic sequestration remains one of the most frequent and life-threatening events(Duminuco et al., 2024;Bokhari et al., 2025). It is particularly prevalent in childhood, typically manifesting below five years, In the regions where hemoglobinopathies are common, with appropriate care, many individuals with SCA can survive into adulthood without significant functional impairment of the spleen(Brousse et al., 2024)(Brousse et al., 2014). Early recognition of splenic sequestration is critical. Rapid pooling of blood within the spleen can precipitate severe hypovolemic shock, rendering prompt diagnosis and intervention essential to prevent morbidity and mortality (Ballas et al., 2012). Yet, in clinical practice, distinguishing splenic sequestration from other common causes of acute abdominal pain in patients with SCD can be challenging, especially when comorbid processes precede overt signs of acute splenic failure. While confirmation typically relies on complete blood counts, imaging, and Doppler ultrasonography, clinicians often must initiate decision-making before these results are available(Solomon et al., 2022)(Suttorp & Classen, 2021) .The aim of our study is to determine the prevalence of splenic sequestration in patients with sickle cell disease.

2. Materials and Methods

2.1. Study Subjects

The Scientific and Ethical Committee of Karbala University College of Medicine examined and approved this study, (Reference number: 2023HU7). We conducted a retrospective cross-sectional study of 101 patients with sickle cell disease who attended the Hereditary blood disease Center in Karbala Province between August 2023 and October 2024.Data were collected using a structured questionnaire with three sections. The first captured socio demographic characteristics (age, sex, blood group, parental consanguinity, and age at diagnosis). The second focused on clinical history relevant to splenic sequestration and other complications—including infections, and acute chest syndrome along with the age at first sequestration, episode frequency, and presenting symptoms. The third addressed management practices, covering prior blood transfusions, folic acid use, pain-control strategies, and whether splenectomy had been performed, with its reported consequences. This design provides a comprehensive profile of patients' disease burden, clinical course, and treatment patterns. The study protocol was approved, and written consent was obtained from the University of Karbala's/ College of Medicine-Famil department. Data were first curated in Microsoft Excel and then analyzed in IBM SPSS Statistics, version 28. Variables were prespecified as continuous (e.g., age, transfusion frequency) or categorical (e.g., sex, splenectomy status). Continuous variables are presented as mean ± standard deviation, and categorical variables as counts and percentages. Associations between categorical variables were assessed using Pearson's chi-square (χ^2) test. Statistical significance was defined as two-sided $p < 0.05$. This analytic framework enabled a clear characterization of clinical features and management outcomes in patients with sickle cell anemia who experienced splenic sequestration.

3. Results

A total of 101 patients diagnosed with sickle cell disease were included in the study, with a mean age of 7 ± 2.37 years. The majority of patients (63.4%, n=64) fell within the 7–10 years age group. Males constituted (56.4%, n=57) of the study

population. Notably, (65.3%, n=66) of the patients had positive parental consanguinity. Regarding blood group distribution, (40.6%, n=41) of patients had blood group O. There was no significant difference between splenic sequestration and sociodemographic characteristics as shown in Table1.

Table1: Association Between Splenic Sequestration and Sociodemographic Characteristics.

Variable	Categories	Frequency	P value
Age group	1–3	10 (9.9%)	0.512
	4–6	27 (26.7%)	
	7–10	64 (63.4%)	
Gender	Male	57 (56.4%)	0.081
	Female	44 (43.6%)	
Parenteral consanguinity	Negative	35 (34.7%)	0.849
	Positive	66 (65.3%)	
Blood group	A	25 (24.8%)	0.892
	AB	3 (2.9%)	
	B	32 (31.7%)	
	O	41 (40.6%)	

Table2 shows that splenic sequestration occurred in 23(22.8%) from 101of patients with sickle cell disease. The majority of patients who experienced splenic sequestration were diagnosed between the ages of 1–3 years (78.3%, n=18). Diagnosis between 4–6 years was less common in both groups, with (13.0%, n=3) in the splenic sequestration group. Very few patients were diagnosed between 7–10 years, accounting for (8.7%, n=2) . The association between age at diagnosis and splenic sequestration was not statistically significant (P = 0.33).

Table2: Association Between Splenic Sequestration and Age At Diagnosis (N=101)

Variable	Categories	Values	Splenic sequestration		P value
			No	Yes	
Age group at diagnosis	1–3	n	50	18	0.33
		%	64.1%	78.3%	
	4–6	n	22	3	
		%	28.2%	13.0%	
	7–10	n	6	2	
		%	7.7%	8.7%	
	Total	n	78(77.8)	23(22.8)	
		%	100.0%	100.0%	

Table3 presents the relationship between age group and the frequency of splenic sequestration episodes. Although the association was not statistically significant (p= 0.6), patients aged 7–10 years showed the highest proportion of frequent sequestration (53.8%), followed by those aged 4–6 years (38.5%). Only 7.7% of patients with frequent sequestration were in the 1–3 years group. While older age groups appear more represented among those with recurrent episodes,

Table3: Association Between Recurrent Splenic Sequestration and Age Group (N=23).

Age Group (years)	No Frequent Sequestration n (%)	Frequent Sequestration n (%)	Total	P value
1–3	1 (50.0)	1 (50.0)	2	
4–6	3 (37.5)	5 (62.5)	8	
7–10	5 (38.5)	8 (61.5)	13	**0.900**
Total	**9 (39.1)**	**14 (60.9)**	**23**	

Table4 shows a significant association between the type of sickle and the occurrence of splenic sequestration (p = 0.027). All cases of splenic sequestration occurred in patients with identifiable splenic pathology. The majority (16.83%) had SS-type, while 3.96% had SD-type, and only 1.98% had SB-type. This highlights that SS-type is the most strongly associated with clinical sequestration episodes.

Table4: Association Between Sequestration and Type of Sickle Cell Disease (N=101)

Sickle Cell Disease Subtype	No Splenic Sequestration n (%)	Splenic Sequestration n (%)	Total	P value
Sβ-thalassemia	22 (91.7)	2 (8.3)	24	-
SD	20 (83.3)	4 (16.7)	24	-
SS	36 (67.9)	17 (32.1)	53	**0.027**
Total	**78 (77.2)**	**23 (22.8)**	**101**	

Table5 presents data indicating that 91.3% of patients who experienced splenic sequestration did not have a splenectomy, whereas 8.7% did, with a no significant association (P = 0.06). Analysis by sickle cell disease type showed that all patients who underwent splenectomy had the SS type, while no splenectomies were performed among patients with SB or SD types; this association was not statistically significant (P = 0.138). Frequent sequestration occurred in 52.2% of patients without splenectomy and in 8.7% of those with splenectomy, also without significant association (P = 0.32).

Table5: Association Between Splenectomy and Other Variables (N=23).

Variable	Category	No Splenectomy n (%)	Splenectomy n (%)	P value
Splenic sequestration	**Yes**	21 (91.3)	2 (8.7)	0.060
Sickle cell disease subtype	**Sβ-thalassemia**	2 (8.7)	0 (0.0)	0.138
	SD	4 (17.4)	0 (0.0)	-
	SS	15 (65.2)	2 (8.7)	-
Frequent splenic sequestration	**Yes**	12 (52.2)	2 (8.7)	0.320
Data are presented as number (percentage). P values were calculated using Fisher's exact test (or Chi-square test, as appropriate).				

Table6 shows the association between splenic sequestration and both hydroxyurea use and regular blood transfusion. There was no statistically significant association between splenic sequestration and hydroxyurea use (p = 0.378). from 101 patients, 23 patients received hydroxyurea, six from 23 (26.1%) had splenic sequestration. In contrast, a highly significant association was found with regular blood transfusion (p = 0.001). from 101 patients, 23 patients received blood transfusion, 18 out of 23 patients with splenic sequestration received blood transfusion.

Table6: Association Between Splenic Sequestration and Preventive Measures.

Variable	Category	No Splenic Sequestration n (%)	Splenic Sequestration n (%)	P value
Hydroxyurea therapy	Yes	17 (73.9)	6 (26.1)	**0.378**
Regular blood transfusion	Yes	5 (21.7)	18 (78.3)	**0.001**
Data are presented as number (percentage). P values were calculated using the Chi-square test or Fisher's exact test, as appropriate.				

4. Discussion

In this study there was no statistically significant association was found between splenic sequestration and sociodemographic information of patients such as age, gender, consanguinity, or blood group. This result agrees with Khaled, et al 2020, which reflects age may affect disease severity, other demographic factors are poor predictors of splenic complications(Khaled et al., 2020). Most patients who experienced splenic sequestration were diagnosed between 1–3 years old (78.3%), though not statistically significant , this finding agreed with (George, et al 2021) which observed early splenic involvement in SCD(George et al., 2021 (Zayed et al., 2025) and (Bokhari et al., 2025), but disagree with (Allali et al., 2024) who experience splenic sequestration later onset due to use of hydroxyurea in early life which improved splenic function and perfusion . In our study percentage of splenic sequestration was 22.77 % , disagree with ((Ladu et al., 2021, Zayed et al., 2025 and Bokhari et al., 2025), 10%, 14.7% and 8.3% respectively may be due to the type of gene

haplotype inheritance. Patients aged 7–10 years had the highest proportion of recurrent splenic sequestration, although the result was not statistically significant. This observation aligns with Ali, et al 2024 ,which reported recurrent episodes in older children due to persistent splenic function (Muayad Mohammed Ali et al., 2024) .There was a statistically significant association between SS-type sickle cell disease and the occurrence of splenic sequestration, our result agrees with George, et al 2021and Roshdy, et al 2024 , both noted SS genotype are most susceptible to early splenic complications than other phenotype (George et al., 2021and Roshdy et al., 2025). But disagree with Ladu *et al* ,2021 and Allard P et al 2024 who revealed occurrence of sequestration occurs more in S/B0disease may be due to no or very small amount of Hb A in this phenotype(Ladu et al., 2021 and Allard et al., 2024).The most reported complications following splenic sequestration were bacteremia, painful crisis, and gallstones. This finding is consistent with (Desai, et al 2023, Roshdy, et al 2024 and Roshdy et al., 2025). Splenectomy was not significantly associated with sequestration and recurrent episodes , with only 8.7% of recurrent cases undergoing splenectomy ,these results agree with (Muayad Mohammed Ali et al., 2024 and Roshdy et al., 2025). There was no significant association found between hydroxyurea use and sequestration ,our study disagree with ,(Allali et al., 2024), while a highly significant association was observed between sequestration and regular blood transfusion ,this result agrees with (John et al., 2020,Celello & McKinney, 2024and Bokhari et al., 2025).

Conclusion

Splenic sequestration is a common and serious complication in children with sickle cell disease, particularly affecting those with the SS genotype. Most cases of splenic sequestration occurred between ages 1–3. No significant association was found between splenic sequestration and sociodemographic factors such as age group, gender, blood group, or parental consanguinity. Regular blood transfusion was significantly associated with splenic sequestration. Hydroxyurea use was not associated with reduced risk of splenic sequestration in this study. Infection is the most risk factors associated with sequestration.

Conflicts of Interest

There is declare no conflict of interests.

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