Indications for and complications of transfusions in patients with sickle cell disease

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Abstract

Background*:* sickle cell disease typically has acute complications that may require transfusions. Although there is a significant prevalence of the disease in Colombia, the characterization of transfusion treatment has not been studied much.

Method*:* this was a descriptive, cross-sectional study at Hospital San Vicente Fundación, in Medellín, Colombia, from January 2011 to December 2023. Patients over the age of 15 with sickle cell disease and complications were included.

Results*:* a total of 106 patients were included, with an average age of 30.9 years, and a predominance of the SS phenotype (90.6%). The main complications were vaso-occlusive crisis (VOC) (74.5%), acute symptomatic anemia (ASA) (61.3%) and acute thoracic syndrome (ATS) (32.1%). Altogether, 85% received transfusions, mainly ordered to treat those with ASA, hepatic sequestration, splenic sequestration, multiple organ failure (MOF) and ATS. Red cell exchange was only performed in seven patients. There were two transfusion reactions, and iron overload affected 36.8% of the patients. The overall mortality was 21.7%, according to the Social Security registers.

Conclusion*:* the study was remarkable for the high frequency of transfusions in patients with sickle cell disease, especially due to ASA, and red cell exchange was infrequently used, even in complications like ATS, MOF and stroke. The results provide the basis for future studies to evaluate the impact of the infrequent use of erythrocyte exchange and iron overload complications, especially their relationship with the high mortality found. **(Acta Med Colomb 2025; 50. DOI: https://doi.org/10.36104/amc.2025.3785).**

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Introduction

Sickle cell disease is one of the most important hemoglobinopathies worldwide, with a large public health impact due to the high morbidity and mortality it causes in affected patients (1).

The geographic regions with the highest burden of this disease are sub-Saharan Africa, South Asia, North Africa and the Middle East, which concentrate most of the new cases and the highest associated mortality. In the Americas, Colombia is one of the countries with the highest number of affected newborns (2). The prevalence of the hemoglobin S mutation in the black population is estimated at 10-11.9%. According to the biweekly national epidemiological report, the national prevalence in 2016 was 0.07 per 100,000 inhabitants, increasing to 0.32 in 2017 (3–5).

The most important acute complications include acute chest syndrome (ACS), painful vaso-occlusive crises (VOC), cerebrovascular accidents (CVAs), symptomatic acute anemia (SAA), priapism, and hepatic and splenic sequestration, among others (6). Red blood cell transfusion is one of the pillars of treatment, and therefore more than 90% of adults with this condition will receive at least one transfusion in their lifetime. The usefulness of transfusion treatment is partially explained by improved blood flow due to a reduced number of sickled cells, as well as its association with reduced endothelial injury and inflammatory damage (7, 8).

However, red blood cell transfusions entail the risk of adverse events like alloimmunization, hemolytic reactions, acute hyperhemolysis and iron overload. In addition, as with any blood product, there are risks of anaphylaxis, minor allergic reactions and pulmonary complications like transfusion-related acute lung injury (TRALI) or transfusion-associated circulatory overload (TACO), as well as transfusion-related sepsis (9, 10).

There are few studies in Colombia characterizing these patients and, to date, none have described the variables related to transfusion therapy, despite being a common treatment strategy for this disease. Therefore, the objective of this study was to characterize the complications derived from sickle cell disease and transfusion treatment in patients treated at a quaternary care hospital in Medellín, Colombia.

Method

Study design and setting

This was a descriptive, observational cross-sectional study that collected retrospective data on patients with sickle cell disease seen between January 2011 and December 2023 at Hospital San Vicente Fundación, Medellín, Colombia.

Population

Patients over the age of 15 with a diagnosis of sickle cell disease were identified based on the ICD-10 codes (D570, D571, D572 and D578, with or without D560, D561, D563, D568 and D569) in their electronic charts. They were then reviewed to verify a documented history or hemoglobin electrophoresis report as well as hospital admissions for disease complications. Patients with sickle cell trait (heterozygous patients with hemoglobin A and S without beta thalassemia) and pregnant patients were excluded.

Variables

Clinical variables like age at diagnosis, phenotype and the main laboratory data available at the time were considered. Acute disease complications were also included, along with the relevant clinical and paraclinical variables for each event as well as the use of hydroxyurea and the need for transfusion therapy.

Transfusions were categorized by type (manual, manual or automated erythrocyte exchanges), the number of red blood cell (RBC) units required, and the presence of complications or adverse reactions. In addition, screening for, diagnosis and treatment of iron overload was examined, as well as vaccine schedule adherence (pneumococcus, influenza and meningococcus), the total number of complications and hospital admissions, mortality and the Charlson index to evaluate the burden of comorbidities (see supplementary appendix for the complete definition of the variables).

Source of data

The data were obtained from the patients' medical charts, as well as diagnostic aids and nursing records. The patients' vital status was verified in the Unique Database of Enrollees (BDUA, in Spanish) of the General Health Social Security System. All collected data were unified and digitized in a Microsoft Excel database (Microsoft 365 licensed to Universidad de Antioquia).

The first event recorded in the patients' medical chart was used for the clinical and paraclinical variables of each type of complication. For the transfusion-related variables, all documented events were considered.

Sample size

A convenience sample was taken, without calculating the sample size beforehand. The number of patients was determined according to the number of patients who consulted at the hospital and met the inclusion and exclusion criteria during the period evaluated by the study.

Quantitative variables

The normality of the distribution of each numerical variable was evaluated with the Kolmogorov-Smirnov test. Medians were reported for variables without a normal distribution to indicate the central value of the group of data.

The Charlson comorbidity index was grouped dichotomously as less than or equal to 1, or greater than or equal to 2. Furthermore, when expressing each of the acute complications, the proportion and absolute number refer to the patients who had that complication at least once during the study period.

Statistical analysis

Absolute and relative frequency distributions were used for the statistical analysis of qualitative variables in each of the categories. Measures of central tendency, like mean and standard deviation (SD) or median and interquartile range (IQR) were used for quantitative variables, according to their distribution. STATA-14 software (licensed to Universidad de Antioquia) was used.

During the course of the study, certain variables were found to have missing data, due to lack of information or records in the medical chart. A simple imputation approach was used to handle this missing data, assigning a value equal to the mean of the available data.

Ethical aspects

The study protocol was submitted to and approved by the research ethics committee at Hospital San Vicente Fundación, Medellín, Colombia. It was determined that the study did not involve any invasive procedure or special risk for the patients. The study ensured the protection of patients' privacy, and identifying data were not revealed.

Results

Out of the 913 records available, 551 patients were evaluated to determine their eligibility, as they were 15 years old or older. Of these, 274 did not meet the inclusion criteria or had an exclusion criterion, and 170 had no record of a hospital admission related to their sickle cell disease. Ultimately, 106 patients were included in the database (Figure 1).

The patients' mean age at the time of data collection was 30.9 years (SD 12.3), while the median age at diagnosis was three years ((IQR 0.75–7, with 26.4% missing data). Sixty-four patients were male (60.4%) and the main sickle

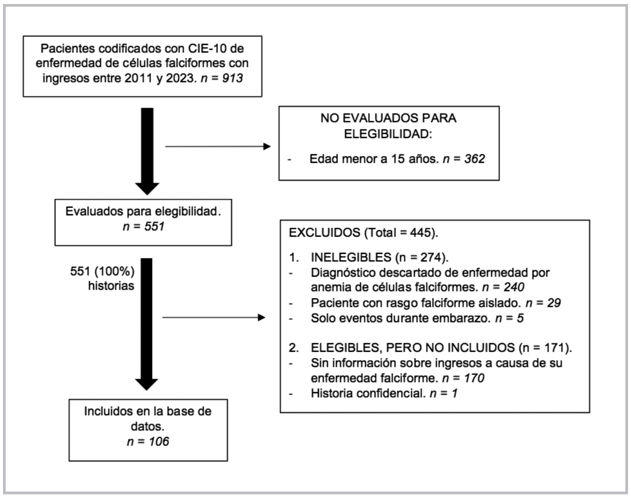

Figure 1. *Patient selection flowchart.*

cell disease phenotype was SS, accounting for 90.6% of the patients. The phenotype was documented by a hemoglobin electrophoresis report in 54.7% of the patients, while for the remaining patients it was assumed from what was written in the medical chart.

Other variables of interest, like the immunization status, Charlson index and median of complications, hospitalizations and RBC units transfused per patient are described in Table 1.

The main sickle cell disease complication was VOC in 74.5% of the patients, followed by SAA in 61.3% and ACS in 32.1%. Table 2 describes the frequency of all complications and their main clinical and paraclinical characteristics. The main reported cause of SAA was hemolysis due to sickle cell crisis, in 95.4% of these patients, 4.6% of cases were due to hepatic sequestration, 3.1% were due to splenic sequestration, and 1.5% were due to bone marrow aplasia with a negative parvovirus B19.

Regarding treatment, more than 70% of the patients were on hydroxyurea when the complication occurred, and 85% underwent transfusions during their disease. The three most common indications for transfusion were, in descending order, SAA, VOC, and ACS. However, transfusion therapy was a treatment strategy for all complications, and was used most often in SAA, hepatic sequestration, splenic sequestration and multiple organ failure (MOF). Table 3 describes the main indications and types of transfusion therapy, the mean RBCs used and the frequency of prior hydroxyurea treatment, desegregated for each complication.

Only one patient was on secondary prevention with chronic transfusions when the second ischemic cerebrovascular event occurred (two years after the first event) and none were receiving primary prevention.

On the other hand, of the patients with MOF, one third resolved with transfusion therapy while the rest died (including one patient who received exchanges). In addition, five patients (4.7%) received a preoperative transfusion at

Table 1. *General and demographic variables of patients with sickle cell disease treated from 2011 - 2023.*

Variable	Patients (n 106)
Mean age (SD)	30.9 (12.3)
Males % (n)	60.4% (64)
Median age at diagnosis (IQR)	3 (0.75-7)
Hgb phenotype %	
SS	90.6
S-beta thalassemia	5.7
SC	3.8
Median of total complications per patient (IQR)	3 (2-6)
Median of total hospitalizations per patient (IQR)	2.5 (1-5)
Median of total units of RBCs transfused per patient (IQR)	4 (2-10)
Immunizations %	
Pneumococcus	8.4
Influenza	7.5
Meningococcus	2.8
Charlson index %	
≤ 1	90.6
≥ 2	9.4
Overall mortality % (n)	21.7 (23)
Median time in months between the last recorded complication and death (IQR)	10 (0.5-37.5)

SD: standard deviation; IQR: interquartile range; Hgb: hemoglobin; RBCs: red blood cells.

some point (hip replacement due to avascular necrosis, hysterectomy due to myomatosis, chest injury, right arthrotomy and cholecystectomy) with an average of two units of RBCs transfused for this reason. The mean preoperative hemoglobin ranged from 7.2 to 8 g/dL, with a mean postoperative hemoglobin of 7.5 g/dL, without significant perioperative bleeding reported in any of the procedures.

Thirty-one patients (29.2%) had complications other than the ones described above. These included pulmonary hypertension as the most frequent (51.6%), followed by pulmonary embolism (19.4%), chronic kidney disease (19.4%), avascular necrosis of the hip (16.1%), priapism (16.1%), heart failure syndrome (9.7%) and cardiomyopathy (9.7%). These complications only motivated simple transfusions on four occasions, with an average of four units of RBCs.

Despite all the transfusions recorded, only two transfusion reactions were documented, which were TACOs. The signs and symptoms in these patients were dyspnea and peripheral edema, with an average length of hospitalization of 21 days, and no deaths related to the transfusion reactions. The indications for transfusion in these cases were ACS and VOC.

However, iron overload was frequent, occurring in 39 patients (36.8%), 15 of whom were managed with iron chelation therapy. Screening for iron overload was done in most patients (no information in 31.1%), with ferritin (65.1%) or magnetic resonance imaging (3.8%).

Table 2. *Frequency and main characteristics of sickle cell disease complications.*

Variables	VOC (74.5%, n 79)	SAA (61.3%, n 65)	ACS (32.1%, n 34)	CVA (5.6%, n 6)	HS (5.7%, n 6) and SS (1.9%, n 2)	MOF (5.7%, n 6)
Mean events per patient (SD)	4.2 (5.2)	2.5 (2.1)	1.7 (1.3)	1.2 (0.4)	1 (0)	1 (0)
Mean Hgb S (SD) *	71.3% (20.1)	76.3% (20.8)	65.9% (18.8)	71.4% (29.6)	62.8% (16.9)	73.8% (23.8)
Mean Hgb (SD)	8.5 g/dL (3.1)	6.6 g/dL (1.6)	7.5 g/dL (1.6)	12 g/dL (3)	6.6 g/dL (2.3)	5.8 g/dL (1.8)
Mean hematocrit (SD)	23.1% (6.3)	19.4% (4.6)	21.8% (5.1)	34.6% (9.5)	18.6% (7.4)	20% (11.5)
Mean WBCs (SD)	14,997 /uL (5246)	17,965 /uL (11611)	20,344 /uL (13578)	10,830 /uL (602)	13,512 /uL (4,719)	16,150 /uL (8,793)
Mean RPI (SD) **	8.6 (4.9)	7.7 (5.5)	6.1 (4)	6.5 (3.5)	9.2 (5.1)	12.2 (9.6)
Median total/indirect bilirubin (IQR)	3.8/2.5 mg/dL (1.4-5.8/1.4-4.3)	4.1/2.4 mg/dL (2.5-6.9/1.2-4.5)	3.7/2.5 mg/dL (2.3-5.9/1.2-4.4)	4.4/3.7 mg/dL (3.5-5.2/2.8-4.6)	8.2/4.7 mg/dL (5.9-21.8/2.1-9)	29.5/8.1 mg/dL (16.4-43.1/6-11)
Median LDH (IQR)	652 U/L (485-863)	678 U/L (554-868)	726 U/L (477-1150)	640 U/L (611-669)	1382 U/L (868-1555)	1984 U/L (1062-3973)
Mean PO2/FiO_2 (SD)	N/A	N/A	270 mmHg (104)	N/A	N/A	N/A
Mean PCO_2 (SD)	N/A	N/A	36 mmHg (10.1)	N/A	N/A	N/A
Median platelets (IQR)	N/A	N/A	N/A	N/A	90 x 1,000/uL (48.5-141)	32.5 x 1,000/uL (24 - 51.5)
Median ALP (IQR)	N/A	N/A	N/A	N/A	N/A	269 U/L (110-518)
Median AST (IQR)	N/A	N/A	N/A	N/A	N/A	3,167 U/L (822-5,773)
Median ALT (IQR)	N/A	N/A	N/A	N/A	N/A	1,079 U/L (465-1,945)
Mean creatinine (SD)	N/A	N/A	N/A	N/A	N/A	2.7 mg/dL (1.6)
Clinical signs and symptoms	Pain in: Extremities 35.4% Lumbar back 22.8% Abdomen 15.2% Sternum 13.9% Hip 12.7%	N/A	Dyspnea 47.1% Chest pain 35.3% Fever 11.8% Cough 5.9%	Hemiparesis 66.7% Headache 33.3%	Abdominal pain 50% Jaundice 25% Fever 12.5% *Shock* 12.5%	Organ dysfunction: Lung 100% Liver 83.3% Kidney 66.7% CNS 50%
Radiological findings	N/A	N/A	Infiltrates on chest x-ray: Interstitial 44% Alveolar 44% Mixed 12%	Brain MRI: Ischemic areas 100%	Tomography: Hepatomegaly 50% Liver infarction 25% Splenomegaly 12.5% Splenectomy 12.5%	N/A

** The missing data is relevant, which amounted to 55.7%, 67.7%, 47.1%, 50%, 37.5% and 33.3% for VOC, SAA, ACS, CVA, HS/SS and MOF, respectively.*
*** The missing data is relevant, which amounted to 30.4%, 26.2%, 35.3%, 66.7%, 37.5% and 33.3% for VOC, SAA, ACS, CVA, HS/SS and MOF, respectively.*
VOC: veno-occlusive crisis; SAA: symptomatic acute anemia; ACS: acute chest syndrome; CVA: cerebrovascular accident; HS: hepatic sequestration; SS: splenic sequestration; MOF: multiple organ failure; SD: standard deviation; Hgb S: hemoglobin S; Hgb: hemoglobin; WBCs: white blood cells; RPI: reticulocyte production index ;IQR: interquartile range; LDH: lactate dehydrogenase; PO2/FiO2: partial pressure of oxygen/fraction of inspired oxygen; PCO2: partial pressure of carbon dioxide; ALP: alkaline phosphatase; AST: aspartate aminotransferase; ALT: alanine aminotransferase; MRI: magnetic resonance imaging; CNS: central nervous system.

Overall mortality was 21.7%, which in absolute numbers corresponds to 23 out of 106 patients. The median time between the last recorded complication and death was 10 months, while the median between the last recorded transfusion and death was 360 days (IQR 40-1,132). However, it is worth noting that of the 23 patients who died, the perimortem care report was only obtained for six, four of whom died from MOF and two from ACS.

Discussion

This study described the main characteristics of transfusion therapy in patients with sickle cell disease complications. We found that the most frequent indication for transfusion was SAA and that the vast majority of patients had undergone transfusion therapy during the course of their disease. In addition, we found a low record of transfusion reactions, but a significant prevalence of iron overload.

The vast majority of the patients had a homozygous SS phenotype, followed by double heterozygous SC and, finally, heterozygous S-beta thalassemia, which is similar to what has been reported in other populations in South America, Europe and countries like Nigeria and Senegal (11-14). The latter countries are relevant, as hemoglobin S haplotypes are mostly of African origin, mainly Bantu and from Senegal,

Table 3. *Description of transfusion therapy and prior treatment with hydroxyurea.*

	Simple transfusions % (n)	Manual exchanges % (n)	Automated exchanges % (n)	Median RBCs used per patient (IQR)	Being treated with hydroxyurea % (n)
SAA (n 65)	100 (65)	0 (0)	0 (0)	4 (2-8)	73.7 (48)
HS/SS (n 8)	100 (8)	0(0)	0 (0)	6 (3-10)	87.5 (7)
MOF (n 6)	83.3 (5)	0 (0)	16.7 (1)	12 (7-16)	83.3 (5)
ACS (n 34)	82.4 (28)	2.9 (1)	2.9 (1)	2.5 (2-5)	70.6 (24)
VOC (n 79)	48.1 (38)	1.3 (1)	2.5 (2)	1 (0-4)	74.7 (59)
CVA (n 6)	33.3 (2)	16.7 (1)	0 (0)	1 (0-8)	50 (3)

SAA: symptomatic acute anemia; HS: hepatic sequestration; SS: splenic sequestration; MOF: multiple organ failure; ACS: acute chest syndrome; VOC: vaso-occlusive crisis; CVA: cerebrovascular accident; RBCs: red blood cells; IQR: interquartile range.

with significant associations between the SS phenotype and a greater need for transfusions in these populations (3, 14).

Most of the patients in our study were diagnosed with the disease before the age of seven. This contrasts with what Pinto et al. reported in 1991 in a group of patients in the city of Medellín, in whom 50% of the diagnoses occurred after age 10 (15). This difference could be due to a greater availability of diagnostic tests for hemoglobinopathies in children and better access to social security today (3). However, the lack of newborn screening remains a gap to be filled.

In a more recent study in Medellín, Martínez et al. found an age at diagnosis similar to our study's, with a median of three years and a male majority (16). They also reported that only 19% of the patients had received transfusions in the last year, which contrasts with our study in which we found that 85% of the patients had received transfusion therapy at some point. In this regard, we are more similar to other countries in the region, like Brazil, where 82.6% of a group of 122 patients had received transfusions (11). However, in other parts of the world, such as France, Nigeria and Senegal, the frequency of transfusion therapy is more heterogenous, at 78.4, 39.1 and 28.5%, respectively, possibly related to the baseline characteristics of the included populations; for example, the high representation of adults in the French studies (12-14).

We found that almost all the transfusions were simple, and RBC exchange was only done in seven patients (three manual and four automated exchanges). These exchanges were for ACS, MOF, CVA and VOC, and were most frequently used for VOC, despite the fact that, according to the current guidelines, VOC in itself is not a standard indication for urgent transfusion or exchange. On the other hand, exchange is more consistently recommended for the other complications, which makes its lack of use noteworthy (for example, five out of six patients with MOF were only managed with simple transfusions) (7, 17). One possible explanation for the low frequency of exchanges in patients with ACS, the third most frequent complication, may be that most did not have severe or rapidly progressive disease and hemoglobin levels were not high (a mean of 7.5 mg/dL).

Most simple transfusions were done for SAA, followed by VOC, likely due to these being the most frequent complications, especially the second one. When collecting data, care was taken in assigning each transfusion indication, considering that some patients with VOC had another concomitant complication that actually justified the transfusion. Even so, we found that almost half of the patients with VOC were managed with simple transfusions, despite average hemoglobin levels of 8.5 g/dL. One of the reasons mentioned was VOCs with pain that was not controlled by the initial treatment. A similar finding was reported in a pediatric study in Yemen by Al-Saqladi et al., in which the most common indications for transfusion were SAA (41.1%), VOC (13.8%) and ACS (8.7%) (18).

The main complications receiving transfusion therapy were SAA, hepatic and splenic sequestration, MOF and ACS. This was similar to the findings of a Jamaican study by Thame et al. (19), in which patients with SAA (mostly due to aplastic crises and splenic sequestration) and ACS were the ones who received the most transfusions. We also found great similarity in the type of transfusion, as, out of 464 episodes with an acute indication, there were 460 simple transfusions and only four exchanges. In a study by Seck et al. in Senegal (14), the main indication for transfusion was SAA and, unlike our study, the second was lower extremity ulcers (50 and 15.6% of transfusions, respectively). In this study, most were simple transfusions (95.7 vs. 4.3% exchanges). It is noteworthy that the latter complication was not reported in our study population.

In our study, we only found two reports of transfusion reactions, which were TACOs. Comparing this with other studies, Thame et al. (19) only described two transfusion reactions (allergic reaction and non-hemolytic fever), while Seck et al. in Senegal (14) found 20 incidents in 339 transfusions performed (mostly mild allergic reactions). A multicenter study reviewing 4,857 transfusions for any indication found that 1.1% were associated with serious reactions (mostly TACO), while mild reactions like allergy and non-hemolytic fever were associated with 0.29 and 0.62% of the transfusions, respectively (20). Based on this, it is unclear whether our low frequency of transfusion reactions is due to a low incidence or under-reporting.

Iron overload is a frequent complication in patients with sickle cell disease, especially those who have received 11-20 transfusions, as described in a survey of doctors in different areas of Europe, Latin America, United States, the Middle East and Africa, in which they reported that 51% to 75% of their patients had iron overload, with serum ferritin measurement being the main assessment method (21). However, it was relatively low in our study (36.8%), despite relatively high screening with the same method.

We found a 21.7% global mortality rate during the study period, but only 5.7% could be attributed to sickle cell disease complications through availability of the medical chart at the time of death. In the study by Thame et al. there was a 19.3% mortality rate among patients who had received transfusions, with a median of 1.3 years between the last transfusion and death (19), similar to our findings. It is interesting to note that, in an American cohort in which 109,783 sickle cell disease hospitalizations were reviewed (28,300 with transfusions and 81,483 without), a relationship was found between mortality and events in which transfusions occurred, compared to those without transfusions (0.43 vs. 0.28%, *p* 0.007) (22).

Our study had some limitations. First, we evaluated the patients' complications and, based on these, we identified which were managed with transfusion therapy. This approach differs from that of other studies which have initially analyzed transfusion episodes and, based on these, have determined their indications, which can make it difficult to compare the results. Second, since this was a single-center study, we cannot determine if the patients had additional complications or required transfusions at other institutions. Furthermore, since our hospital is a reference center, there could be a selection bias, as we are likely to have received more complex cases or cases with more serious complications, which could limit its representativeness of the general population with sickle cell disease. Third, due to the study's retrospective nature, there may be information bias in the data collected.

Despite these limitations, our study has important strengths. We included a significant adult population with a broad representation of diverse disease complications, which contributes valuable information to a field with a scarcity of data. Furthermore, our findings provide evidence of the low frequency of transfusion reactions and high mortality in this population. This supports the need to analytically evaluate the potential factors associated with each outcome, as well as the impact of complications despite the use of hydroxyurea, especially considering the unavailability of therapeutic alternatives in our setting.

Conclusions

There was a high rate of transfusions in patients with sickle cell disease, with SAA as the main indication for transfusion, and with a notably scant use of RBC exchanges in complications like ACS, MOF and CVA. There was a low frequency of transfusion reactions, although the significant iron overload found in these patients is notable, which underscores the importance of implementing appropriate treatment strategies to mitigate this risk. These findings contribute to understanding the clinical management of patients with sickle cell disease, highlighting the need for a comprehensive approach that considers both the risks and benefits associated with transfusion therapy.

References

1. **Kavanagh PL, Fasipe TA, Wun T.** Sickle Cell Disease: A Review. *JAMA*. 2022; 328(1):57-68.
2. **Thomson AM.** Global, regional, and national prevalence and mortality burden of sickle cell disease, 2000–2021: a systematic analysis from the Global Burden of Disease Study 2021. *Lancet Haematol*. 2023; 10(8):e585–99.
3. **Durán CL, Morales OL, Echeverri SJ, Isaza M.** Beta globin haplotypes in hemoglobin S carriers in Colombia. *Biomédica*. 2012; 32(1):103–11.
4. **Rodríguez WE, Sáenz GF, Chávez MA.** Haplotipos de la hemoglobina S: importancia epidemiológica, antropológica y clínica. *Rev Panam Salud Pública*. 1998; 3:1-8.
5. **Misnaza SP.** Drepanocitosis en Colombia: análisis de la notificación como enfermedad huérfana o rara al sistema de vigilancia en salud pública, 2016 y 2017. *Informe Quincenal Epidemiológico Nacional*. 2018; 23(1): 1-13. Disponible en: www.ins.gov.co/buscador/IQEN/IQEN%20vol%2023%202018%20num%201.pdf
6. **Onimoe G, Rotz S.** Sickle cell disease: A primary care update. *Cleveland Clinic Journal of Medicine*. 2020; 87(1):19–27.
7. **Piel FB, Rees DC, DeBaun MR, Nnodu O, Ranque B, Thompson AA, et al.** Defining global strategies to improve outcomes in sickle cell disease: a Lancet Haematology Commission. *Lancet Haematol*. 2023; 10(8):e633-e686.
8. **Kato GJ, Piel FB, Reid CD, Gaston MH, Ohene-Frempong K, Krishnamurti L, et al.** Sickle cell disease. *Nat Rev Dis Primers*. 2018; 4(1):1–22.
9. **Rollins MR, Chou ST.** Adverse events of red blood cell transfusions in patients with sickle cell disease. *Transfus Apher Sci*. 2022;61(5): 103557.
10. **Ackfeld T, Schmutz T, Guechi Y, Le Terrier C.** Blood Transfusion Reactions—A Comprehensive Review of the Literature including a Swiss Perspective. *J Clin Med*. 2022; 11(10): 2859.
11. **Cesar P, Dhyani A, Augusto-Schwade L, Acordi P, Xerez-Albuquerque C, Nina R, et al.** Epidemiological, clinical, and severity characterization of sickle cell disease in a population from the Brazilian Amazon. *Hematol Oncol Stem Cell Ther*. 2019; 12(4):204-210.
12. **Gerardin M, Rousselet M, Couec ML, Masseau A, Guerlais M, Authier N, et al.** Descriptive analysis of sickle cell patients living in France: The PHEDRE cross-sectional study. *PLoS ONE*. 2021; 16(3):e0248649.
13. **Brown BJ, Jacob NE, Lagunju IA, Jarett OO.** Morbidity and mortality pattern in hospitalised children with sickle cell disorders at the UCH Ibadan, Nigeria. *Niger J Paed*. 2013; 40(1): 34 –39.
14. **Seck M, Tall A, Faye BF, Bah DS, Guéye Y, Sall A, et al.** Evaluation of transfusion practices in sickle cell disease in Senegal: cohort study of 1078 patients with sickle cell disease. *Med Sante Trop*. 2017; 27(4):402-406.
15. **Pinto L, Cuéllar F, Maya LM, Murillo ML, Mondragón MC, Alvarez L.** Anemia de células falciformes en adultos: estudio clínico de 51 pacientes tratados en el Hospital Universitario San Vicente de Paúl. *Acta Med Colomb*. 1991; 16(6):309–316.
16. **Martínez-Sánchez LM, Villegas-Alzate JD, Herrera-Almanza L, Correa-Saavedra MA, Gálvez-Cárdenas KM, Hernández-Martínez A, et al.** Caracterización de pacientes con anemia de células falciformes en Medellín, Colombia. *Med Int Méx*. 2022; 38(5):1012-1018.
17. **Han H, Hensch L, Tubman VN.** Indications for transfusion in the management of sickle cell disease. *Hematology Am Soc Hematol Educ Program*. 2021; 2021(1):696-703.
18. **Al-Saqladi AM, Maddi DM, Al-Sadeeq AH.** Blood Transfusion Frequency and Indications in Yemeni Children with Sickle Cell Disease. *Anemia*. 2020; 2020:7080264.
19. **Thame JR, Hambleton IR, Serjeant GR.** RBC transfusion in sickle cell anemia (HbSS): experience from the Jamaican Cohort Study. *Transfusion*. 2001; 41(5):596-601.
20. **Hendrickson JE, Roubinian NH, Chowdhury D, Brambilla D, Murphy EL, Wu Y, et al.** Incidence of transfusion reactions: a multicenter study utilizing systematic active surveillance and expert adjudication. *Transfusion*. 2016; 56(10):2587-2596.
21. **Vichinsky EP, Ohene-Frempong K; Transfusion Committee.** Approaches to transfusion therapy and iron overload in patients with sickle cell disease: results of an international survey. *Pediatr Hematol Oncol*. 2011; 28(1):37-42.
22. **Sharma A, Dahiya A, Alavi A, Woldie I, Karson J, et al.** Patterns of Blood Transfusion in Sickle Cell Disease Hospitalizations. *Hemato*. 2024; 5(1):26-34.