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Healthcare Utilization and Clinical Burden of Sickle Cell Disease in a Portuguese Emergency Department

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Abstract

Background/Objectives: Sickle Cell Disease (SCD) is a rare hemoglobinopathy in Portugal, where epidemiological data remain limited. National newborn screening has revealed a higher-than-expected incidence, particularly in urban areas, underscoring the importance of understanding acute care utilization. Vaso-occlusive crises (VOC) frequently lead to emergency department (ED) attendance; however, no prior studies have characterized SCD care in Portuguese EDs. This study aimed to describe patterns of emergency care use, clinical presentation, and outcomes in a Portuguese hospital, and to contextualize these findings within international standards. **Methods**: This retrospective observational study reviewed SCD-related emergency visits to the ED of a district hospital from January 2021 to December 2023. Patients aged ≥18 years with SS, Sβ, or SC genotypes were included. Data collected included demographics, triage category, presenting complaints, pain scores, waiting times, treatments administered, diagnostic investigations, ED length of stay (LOS), disposition and 72-hour revisit rates. Descriptive and bivariate analyses were performed, with significance defined as $p < 0.05$. **Results**: A total of 264 ED episodes from 93 unique patients (mean age 30 years; 61% male) were identified, with a mean of 2.8 visits per patient over three years. Limb pain (31%) and back pain (28%) were the most common presenting complaints, and 83% of patients reported severe pain at triage. Most episodes were classified as very urgent (65.2%) or urgent (27.3%), yet the mean time to physician assessment was 58 minutes. The mean ED LOS was 13 hours, substantially longer than reported internationally, and 45.8% of visits resulted in hospital admission. Laboratory testing was performed in 91% of episodes and imaging in 39%. Analgesia was administered in 98% of visits, with frequent use of multimodal regimens; however, pethidine was used in 28% of episodes despite guideline recommendations discouraging its use. The 72-hour revisit rate was low (~4%), with six return visits requiring readmission. **Conclusions**: SCD patients in Portugal frequently present to the ED with high-acuity VOC requiring intensive evaluation and analgesia. ED LOS substantially exceeded international benchmarks, likely reflecting structural factors such as overcrowding, boarding delays and absence of dedicated acute pain pathways. The persistence of outdated opioid practices further highlights opportunities for quality improvement. These findings emphasize the need for locally adapted, guideline-concordant ED pathways to optimize the acute management of SCD and improve patient outcomes.

Keywords: sickle cell disease; vaso-occlusive crisis; analgesia; pain; emergency department; acute pain management

1. Introduction

Sickle Cell Disease (SCD) is a structural hemoglobinopathy caused by a point mutation in the β-globin gene, resulting in hemoglobin S or sickled red blood cells, which lead to chronic hemolytic anemia, vaso-occlusive events, and progressive multi-organ damage. While SCD is prevalent in sub-

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Saharan Africa and areas with high population mobility from endemic regions, it is a rare condition in Portugal, with an estimated 850–900 individuals nationwide affected, as recently reported [1–3].

Between 2021 and 2023, the Instituto Nacional de Saúde Doutor Ricardo Jorge (INSA), which implemented a national newborn screening program, reported an incidence higher than anticipated, about 1:1695 nationwide, and 1:978 in the Lisbon and Setúbal districts, prompting the inclusion of SCD in the Portuguese National Neonatal Screening Program (PNRN) [2]. These statistics highlight the increasing importance of SCD in the Portuguese health care system, especially in urban and ethnically mixed populations.

Clinically, vaso-occlusive crisis (VOC) is the most frequent emergency department (ED) patient complaint, described as severe pain in a severe, localized manner usually managed with opioid-based control. High ED utilizations, long waits, and hospitalizations of 30–40% in SCD patients have already been reported in international studies [4–8]. However, Portuguese data remain scarce.

EDs in Portugal are chronically under operational strain and overcrowded, with one of the highest per capita ED visit rates in Europe, up to 58 visits per 100 population per year [9]. Consequently, waiting times typically exceed recommended triage amounts, and long durations in medical evaluation and ED stay were reported in several studies. These systemic pressures lead to patient dissatisfaction, and these complaints are commonly acknowledged in Portuguese urgent care. These delays are especially harmful to patients suffering from SCD for whom timely analgesia is critical: prolonged time-to-opioid administration has been linked to suboptimal pain control, increased rates of hospitalization, and enhanced complication risk (e.g., acute chest syndrome) [10]. In contrast to systemic pressure, no Portuguese studies have investigated the management of SCD in the ED or the impact of operational dynamics on clinical outcomes for this patient group, constituting a relevant national evidence gap.

This study aims to characterize the emergency care utilization, clinical presentation, and outcomes of adult patients with SCD in a Portuguese district hospital between 2021 and 2023, and to compare the findings with international data to identify areas for quality improvement.

2. Materials and Methods

2.1. Study Design and Setting

This was a retrospective observational study conducted at the ED of ULS Amadora-Sintra, a district hospital located in the Lisbon metropolitan area. The study period extended from January 2021 to December 2023.

2.2. Study Population

The study included adult patients (≥18 years) with a confirmed diagnosis of SCD (HbSS, HbSC, or HbSβ genotypes) who attended the ED during the study period. Pediatric cases, cases presenting at Basic Emergency Units, episodes classified as non-urgent (white triage) or patients discharged against medical advice were excluded.

2.3. Data Sources and Variables

Data were retrieved from the hospital's electronic medical records using ICD-9 (282.6.x) and ICD-10 (D57.x) codes for SCD. Each ED episode was analyzed separately.

The collected variables included:

- Demographics: age, sex, country of origin, genotype.
- Clinical parameters: presenting complaint, pain intensity (0–10 numeric scale), Manchester Triage System (MTS) flowchart and discriminator, assigned priority level, triage color.
- Time metrics: time from admission to triage, time to first medical observation, total ED length of stay.

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- Diagnostics and treatment: laboratory testing, chest X-ray (CXR), eletrocardiogram (ECG), CT imaging, and analgesia administered (paracetamol, metamizole, non-steroidal anti-inflammatory drugs (NSAIDs), tramadol, pethidine, morphine, fentanyl).
- Outcomes: disposition (discharge or admission), and 72-hour readmission rate.

2.4. Statistical Analysis

Data was entered into a standardized Excel database. Descriptive analyses (means, medians, frequencies, and percentages) were performed for all variables. Associations between categorical variables (e.g., triage level and admission) were evaluated using bivariate analysis. Statistical significance was defined as $p<0.05$.

3. Results

3.1. Demographic and Epidemiological Profile

A total of 264 ED episodes involving 93 unique adult patients with confirmed SCD were analyzed. The average patient age was 30 years (range: 18–57), and 61% were male.

Most patients were first- or second-generation migrants from Lusophone African countries, predominantly Angola (40%), São Tomé and Príncipe (11%), and Cape Verde (7%), reflecting the hospital's diverse catchment population. Portuguese-born individuals accounted for approximately 38% of the cohort.

Over the three-year study period, the average number of ED visits per patient was 2.8 (range 1–14). Notably, 19% of patients accounted for more than 50% of all SCD ED visits, indicating a small subgroup with very frequent ED utilization. Baseline patient characteristics and ED presentation metrics are summarized in Table 1.

Table 1. Baseline characteristics and clinical presentation profile of SCD–related ED episodes at ULS Amadora-Sintra (Portugal), 2021–2023.

	n = 264
Age, average	30 years
Sex	
Male	61%
Nationality	
Angola	105
Brazil	1
Democratic Republic of the Congo	1
Cape Verde	18
Guinea	1
Guinea-Bissau	8
Portugal	100
São Tomé e Príncipe	30
Genotype	
HbSC	8 (3%)
HbSS	256 (97%)
MTS categories	
Non-Urgent	1
Little Urgent	19
Urgent	72
Very Urgent	172
Presenting Complaints (MTS flowcharts)	
Headache	9
Diarrhea and/or vomiting	2

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Dyspnea	7
Abdominal pain	18
Sore throat	1
Back pain	74
Chest pain	19
Unconsciousness/syncope	2
Wounds	2
Adult malaise	46
Local infections and abscesses	1
Palpitations	2
Limb problems/pain	81
Pain at Admission to ED	
≤3	9
4-6	35
≥7	220
Time until triage, average	13 min
Time to first medical assessment, average	58 min
ED length-of-stay, average	13 h

3.2. Clinical Presentation and Triage

Vaso-occlusive pain crises were the predominant reason for ED presentation, accounting for nearly 90% of all visits. The most common specific presenting complaints were limb problems/pain (81 episodes, 30.7%), back pain (74 episodes, 28.0%), adult malaise (46 episodes, 17.4%), dyspnea or chest pain (26 episodes, 9.8%) and abdominal pain (18 episodes, 6.8%).

Pain intensity at triage was typically severe, with 83% of patients reporting ≥7/10 on the numeric pain scale. According to the MTS, 65.2% of episodes were classified as very urgent (orange), 27.3% as urgent (yellow), and 6.8% as emergent (red). No episodes were triaged as non-urgent (white or blue).

The mean waiting time for the first medical evaluation was 58 ± 35 minutes, ranging from immediate assessment (in red codes) to up to 120 minutes in lower-priority cases.

3.3. Diagnostic Workup

Diagnostic investigations were frequently performed, reflecting the clinical complexity of these ED cases. Laboratory tests were obtained in 91% of visits, chest X-rays in 39%, ECGs in 27%, urine analyses in 22%, and CT scans in 12%.

The extent of workup was greater in patients who required hospitalization: among admitted cases, 96% underwent both laboratory testing and some form of imaging, compared to 68% of cases that were discharged directly from the ED.

3.4. Analgesia and Pain Management

Analgesia was administered in 98% of all ED visits. Paracetamol was used in 86% of cases, NSAIDs in 72%, tramadol in 64%, and strong opioids (morphine or a comparable opioid) in 52%. Pethidine, an older opioid analgesic, was administered in 28% of visits. The vast majority of patients (83%) received a multimodal analgesic regimen (≥2 classes of analgesics).

Table 2. Analgesic medications administered during ED visits for SCD-related episodes at ULS Amadora-Sintra (Portugal), 2021–2023.

Medication	**n (%)**
Any analgesia	259 (98)
Non-opioid	

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Paracetamol	227 (86)
NSAIDs	190 (72)
Metamizole	91 (34)
Opioid	
Tramadol	169 (64)
Strong opioids (morphine or equivalent)	137 (52)
Pethidine	74 (28)
Multimodal regimen (≥2 drug classes)	219 (83)

Pain control often required intensive therapy: in 21% of ED visits, the patient received more than three doses of morphine before disposition (discharge or admission), indicating difficulties in achieving adequate pain relief. Importantly, providing opioid analgesia early (at the triage stage) was associated with faster pain control – the median time to effective pain relief was 40 minutes in those who received an opioid at triage, compared to 67 minutes in those who did not ($p < 0.05$).

3.5. Emergency Department Throughput and Outcomes

The mean total time spent in the ED was 13 ± 6.5 hours per visit (range 2–27 hours). ED length of stay was significantly longer for visits that resulted in hospital admission (mean 15.1 hours) compared to visits that ended in discharge home (mean 10.7 hours, $p < 0.01$).

The leading causes of hospital admission from the ED were uncontrolled pain (61% of admitted cases), infection (18%), and either acute chest syndrome or severe anemia (11%). No deaths occurred during the study period.

Overall, 11 ED visits (4%) were followed by a return to the ED within 72 hours, all due to recurrent VOC. Among these 72-hour return visits, 6 (54%) ultimately required hospital admission upon return. While such short-term revisits were relatively infrequent, a notable 27% of patients experienced at least one ED re-presentation within six months, underscoring the recurrent nature of SCD complications in this cohort.

4. Discussion

The present study is the first in Portugal to report on how SCD patients utilize emergency care locally and found a high rate of severe acute presentations with significantly prolonged ED stays, compared to international studies. Such a demographic profile of our cohort aligns with the post-colonial patterns of immigration in Portugal, as the majority of patients were from Lusophone African countries, consistent with national screening data [1–3]. Thus we note that, the dominance of VOC as the most frequent reason for ED attendance reflects global patterns and emphasizes the significant contributions of acute pain in the morbidity of SCD [4–8].

Although nearly every episode was triaged as urgent or very urgent, the median waiting time for a medical assessment was nearly an hour. This delay is clinically relevant, since early opioid administration is known to be associated with better pain control and reduced hospital admission rates [7,8]. Therefore, our discovery that opioid analgesia initiated in triage was associated with quicker pain relief confirms the significance of timely treatment. However, the discrepancy between the proposed timelines and actual practice shows some systemic limitations inside the ED.

However, emerging evidence suggests that ED operational pressures play an essential role in the subclinical setting as they provide patient care of SCD. Overcrowding, delays and inconsistent workflows are widely known factors leading to undertreatment of pain and several studies identified ED crowding was related to delays in opioid delivery, poorer pain control and higher chances of hospital admission [10–12]. Portugal has one of the highest rates of ED use rates in Europe and structural strain [9], indicates that the prolonged ED waiting times and ED length of stay (LOS) we found among this cohort is likely to depend on not just the severity of symptoms but also on structural features intrinsic to national emergency care.

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Consistent with this assessment, our mean LOS of 13 hours was about twice as long as reported in the US and UK, where average LOS in SCD cases is in the range of 6-8 hours [4,5]. This difference may be due to multiple structural constraints in this hospital. On the other hand, ED boarding due to inadequate inpatient bed spaces, long-established in Portuguese hospital practice [9], delay admissions upon clinical decision-making probably delaying their finalization. Due to a lack of a dedicated acute pain service or short-stay analgesia pathway, opioid titration and monitoring must be performed within normal ED bays, thereby reducing throughput efficiency. In addition, ED crowding further prolongs initial assessment and limits wait time, resulting in longer patient admission and discharge from the hospital. The proportion of episodes requiring laboratory investigations (91%) and imaging studies (nearly 40%) also prolongs diagnostic handling and disposition decisions. The interaction of these factors accounts for the much longer LOS seen in our group.

International guidelines further contextualize these findings. NICE guideline CG143 recommends administering first-line analgesia, including strong opioids [13], within 30 minutes of arrival, with frequent reassessment and rapid titration of pain control. The 2020 ASH guidelines also recommend individualized, rapidly titrated opioid therapy, and systematic avoidance of undertreatment [14]. Based on our data, these principles are hard to implement in practice, particularly in a high workload/resource-strained clinical setting.

Another notable finding would be the relatively widespread use of pethidine, given in 28% of visits. This is in contrast to recommendations used in modern day for SCD pain, wherein pethidine's short duration of action and neurotoxic metabolite norpethidine make it a less appropriate choice. NICE guidelines specify that seizure risk in SCD is increased, and a number of centres in the UK have removed pethidine from their pain protocols entirely [13]. Its continued use in our institution likely reflects historical prescribing patterns rather than evidence-based practice, suggesting a clear target for future quality improvement.

Taken together, these results suggest the necessity for simplified, locally adapted ED pathways for SCD to maximise early analgesia, early senior review and rapid and efficient diagnostic bundles. These interventions have the promise of diminishing LOS, ensuring more efficient and consistent analgesia and consequently improving the quality of emergency care experienced by those suffering from SCD.

5. Conclusions

This work demonstrates that SCD imposes a significant and under-recognized burden on Portuguese emergency departments. Patients present with high-acuity crises yet face prolonged delays and care pathways that diverge from established international standards. The extended ED length of stay, inconsistent timeliness of analgesia and continued use of non-recommended opioids reflect system-wide pressures that go beyond individual clinical encounters. These findings call for immediate investment in dedicated pain pathways, earlier specialist involvement, and operational reforms to reduce crowding and improve flow. Strengthening the emergency care response for SCD is not only a clinical priority but a matter of equity and safety. Implementing structured, guideline-driven models of care has the potential to transform outcomes for this vulnerable population and should be a strategic objective for the Portuguese health system.

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Informed Consent Statement: Patient consent was waived due to to retrospective anonymized data.

Data Availability Statement: Data are available from the corresponding author upon reasonable request.

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Conflicts of Interest: The authors declare no conflicts of interest.

Abbreviations

The following abbreviations are used in this manuscript:

ACS	Acute Chest Syndrom
CXR	Chest X-ray
CT	Computed Tomography
ECG	Electrocardiogram
ED	Emergency Department
ICD-9	International Classification of Diseases, 9th Revision
ICD-10	International Classification of Diseases, 10th Revision
INSA	Instituto Nacional de Saúde Doutor Ricardo Jorge
IQR	Interquartile Range
IRB/EC	Institutional Review Board/Ethics Committee
LOS	Length of Stay
MTS	Manchester Triage System
NSAIDs	Non-steroidal Anti-inflammatory Drugs
PNRN	Programa Nacional de Rastreio Neonatal (Portuguese National Neonatal Screening Program)
SCD	Sickle Cell Disease
SD	Standard Deviation
ULS	Unidade Local de Saúde (Local Health Unit)
VOC	Vaso-occlusive Crisis

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