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Research Article

Sickle Cell Disease Vaso-Occlusive Crisis in the Emergency Department: A Cross-Sectional Descriptive Study for Physicians Prospective and Approach in Saudi Hospitals

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Abstract

Background: Sickle cell disease is an inherited disorder with a relatively significant prevalence in the kingdom of Saudi Arabia. The management of the condition necessitates compliance to certain guidelines. Our study aims to assess the approach of emergency departments physicians in Saudi hospitals to the presentation of Vaso-occlusive crisis.

Methods: A cross-sectional descriptive study, in Saudi Arabia, through a period of three months, starting from 15th of December 2020 to 15th of March 2021. The data were obtained using a predesigned online questionnaire sheet. Approvals from Ethics and Scientific Committees (UB-RES-2020-0052) of Batterjee Medical College were obtained before the conduction of the study.

Results: Majority of the Emergency department physicians stated that they approached the management of sickle cell patients presenting with painful episodes initially by physical examination 51 (34.2%). Preferred first step for the majority 86 (57.7%) was the administration of intravenous fluids for the aim of maintaining good hydration. 61 (40.9%) stated that they preferred instant administration of analgesia. The most common analgesic administered was morphine 97 (65%).

Conclusion: In our study we found that opioids as morphine are prescribed at an increased rate. Despite the adverse effects being known, especially tolerance, prescription of morphine remains to dominate. We recommend further assessment of parameters to enhance pain assessment in terms of reliability.

Keywords: *Anemia; Sickle Cell Disease; Saudi Arabia; Emergency Physician; Vaso-Occlusive Crisis*

Introduction

Sickle cell disease (SCD) is a hereditary disorder of the red blood cells affecting about 72,000 individuals in the United States of America, and more than 200,000 infants are born yearly with SCD in the continent of Africa. SCD is a common genetic disease in Saudi Arabia. About 2 - 27% of the population are carriers of the sickle cell trait, and up to 1.4% have SCD in some areas, with the highest prevalence noted in the Eastern province (145 cases per 10,000). The cardinal factor that causes SCD to be common in Saudi Arabia is the prevalence of consanguineous marriages (57.7%) [1-3]. The main type of hemoglobin in SCD is Hemoglobin S that causes the red blood cells to be less flexible and get sickled when exposed to certain factors. The triggering factors include hypoxia, infections, stress, and dehydration. The resulting sickle-shaped red blood cells can occlude blood vessels causing the tissues to get hypoxic and the patient to be manifested with painful crisis (vaso-occlusive crisis), which is the most common cause of the recurrent Emergency Department (ED) visits by SCD patients. Other complications of SCD include hemolytic crisis, aplastic crisis, pneumonia, acute chest syndrome, meningitis, stroke, splenic sequestration crisis, hepatic crisis, avascular necrosis, osteomyelitis, priapism, and dactylitis [4,5].

According to the National Heart, Lung, and Blood Institute (NHLBI) guidelines for managing sickle cell pain in the ED, a physician's quick assessment and triage as an urgent case to be evaluated by a physician is essential for patients with acute pain. First, pain should be well-assessed to exclude other underlying complications or causes as infections or cardiopulmonary issues. After ruling out other causes of pain, sickle cell pain should be managed by an analgesic to be administered within 60 minutes of presentation to the ED. The Intravenous (IV) route is the best route to administer the medications. Still, the subcutaneous route can also be beneficial for difficult venous access or to reduce the time to receive the first analgesic dose in a crowded ED. The intramuscular route shouldn't be used as it's very painful and has no additional advantage over subcutaneous administration. Reassessment of the pain should be done every 15 - 30 minutes to respond to analgesics and repeat the doses until the pain is relieved. Nonsteroidal anti-inflammatory drugs (NSAIDs) or acetaminophen can be used to subside mild-to-moderate pain, while persistent or severe pain can be managed with opioids. As SCD patients could have pre-existing liver impairment, physicians should be cautious regarding the doses of acetaminophen and NSAIDs to avoid the development of complications, including liver failure, peptic ulcer, gastritis, and renal failure [6,7].

An additional factor associated with prolonged time to assess and treat SCD patients in the EDs is believed to be that the presenting individuals are drug addicts, although no proof confirms this belief [5]. A study done by Shapiro BS., *et al.* showed that 53% of ED physicians consider that more than 20% of SCD patients are addicts. However, the chance of SCD patients becoming addicted is like that of the general population (2-5%). This belief affects physician's attitudes toward the patients. It can result in undertreatment of pain crisis, as they found that ED physicians with a negative attitude are less likely to follow the guidelines [6,8,9].

Several screening tools can be used to identify patients at high risk to develop opioid addiction so they can be managed carefully, including CAGE-AID questions, Drug Abuse Screening Test (DAST), Opioid Risk Tool (ORT), Addiction Behaviors Checklist and the Diagnostic and Statistical Manual of Mental Disorders, 4th Edition, Text Revision (DSM-IV-TR) criteria [10-12].

Materials and Methods

Study design and setting: A cross-sectional descriptive study, in Saudi Arabia, through a period of three months, starting from 15th of December to 15th of March 2021.

Study population and sampling technique: All ED Physicians living in Saudi Arabia and agreeing to participate in the study were eligible to be recruited in the study. Minimum sample size was calculated considering a level of confidence of 95%, expected prevalence of 50%, and precision of 0.05 and was found to be 150. Total sample collected was 149. To comply with the physical distancing rules in response to the COVID-19 pandemic, recruitment of study participants was done via physical interviews in the hospital setting.

Study tool: The data were obtained using a predesigned online questionnaire sheet. For every participant, the following data were collected (a) Preferred approach to assessment of pain in SCD patients presented with a vaso-occlusive crisis (VOC) (b) Preferred first step in management of SCD patients presented with VOC (c) Time gap between patients' presentation and administration of analgesics to SCD patients presented with VOC (d) Preferred analgesia type prescription for SCD patients presented with VOC(e) Common adverse effects of analgesia identified by ED physicians in patients with SCD.

Ethical considerations: Approval from Ethics and Scientific Committees (UB-RES-2020-0052) of Batterjee Medical College were obtained before the conduction of the study. Informed online consent was obtained from each participant; the aim of the study was clearly explained and the "Agree to participate" icon was a condition before proceeding in responding to the questionnaire items. Data were collected anonymously and the confidentiality of collected data was guaranteed.

Data analysis: The collected data were statistically analyzed using statistical package for social studies (SPSS) version 23 created by IBM, Chicago, IL, USA. Categorical variables were presented as numbers and percentages.

Results

The current study included 149 physicians in some Emergency departments in the Eastern and Western provinces of Saudi Arabia. Any information that may have led to identification of the participating physician or their hospital have been maintained confidential. Upon assessment of the preferred approach to assessment of pain in SCD patients presented with VOC (Table 1), most of the ED physicians stated that they approached the patient initially by physical examination 118 followed by vital signs 109. A second question was asked to all study participants aiming to assess the preferred first step in the management of SCD patients presented with VOC (Table 2). The results showed that the preferred first step for the majority 86 (57.7%) was found to be the administration of intravenous fluids for the aim of maintaining good hydration. Additionally, 61 (40.9%) stated that they preferred instant administration of analgesia. The most prevalent average time between presentation in the ED and administering an analgesic was less than 30 minutes, as reported by 84 (56.4%) of the ED physicians. In contrast, 56 (37.6%) stated that patients with SCD wait for 30 - 60 minutes before receiving any analgesic (Table 3). Detailed questioning on the type of analgesia was asked, and the vast majority stated that the most common analgesic prescribed by ED physicians was morphine 97 (65%) (Table 4). The last question aimed to identify the most common adverse effects of analgesics that SCD patients present with, and the most identifiable one tended to be tolerance 132 as was evident in our study (Table 5).

Preferred Approach	Frequency
Physical Examination	118
Vital Signs	109
History	62
Reticulocyte Count	52
Hemoglobin Level	51

Table 1: *Emergency department physicians preferred approach to assess pain in sickle cell disease patients (Participating physicians were allowed to choose more than one choice).*

Preferred Step	Frequency	Percentage (%)
Good Hydration	86	57.7
Analgesia	61	40.9
Hydroxyurea	1	0.7
Blood Transfusion	1	0.7
Total	149	100

Table 2: *Emergency department physicians preferred first step in the management of Sickle Cell Disease patients with a vaso-oclusive crisis.*

Time Gap (minutes)	Frequency	Percentage (%)
< 30	84	56.4
30 - 60	56	37.6
> 60	9	6
Total	149	100

Table 3: *Time gap between patients' presentation and administration of analgesics to sickle cell disease patients presented with pain.*

Analgesia Type	Frequency	Percentage (%)
Morphine	98	65
Acetaminophen	26	17
Tramadol	13	9
NSAIDs	12	9
Total	149	100

Table 4: *Most common analgesia type prescribed by Emergency department physicians for sickle cell disease patients with a vaso-oclusive crisis.*

Adverse effect	Frequency
Tolerance	132
Tachycardia	25
Respiratory depression	25
Gastrointestinal and Urinary Symptoms	4
Dizziness	1
Addiction	1

Table 5: *Common adverse effects of analgesia identified by emergency department physicians in sickle cell disease patients.*

Discussion

Our study aims to identify and assess the approach to the management of vaso-occlusive crisis (VOC) in patients with SCD in the Emergency departments in Saudi Arabia. We collected data from physicians in multiple EDs of both governmental and private hospitals in the Eastern and Western provinces of Saudi Arabia. We found that out of 149 Physicians, 112 (75.2%) stated that their hospitals have protocols to manage patients with SCD. A previous study reported that efficient protocols in EDs could help decrease ED visits and over-consumption of ED resources. Another study stated that sickle cell pain management protocol in EDs could reduce ED visits and increase outpatient clinic utilization.

About 53 (35.6%) of the physicians stated that the most common complaint that patients with SCD present to the ED with is painful episodes (VOC). This finding was also reported in a study conducted by Koshy M., *et al.* in which they found that 37% of ED visits by the patient with SCD was due to painful episodes, 35% of which had a determining cause and 75% of them needed to be admitted. A descriptive study that studied data from National Hospital Ambulatory Care Surveys (NHAMCS) from 1999 to 2007 showed that pain symptoms were also the main complaint representing 67% of all the complaints that SCD patients presented to the EDs with [13,14].

Physical examination is considered as an indicator to help in pain assessment of patients with SCD by most of the ED physicians (79.2%), followed by vital signs (73.2%) and history (41.6%). According to NHLBI guidelines for managing sickle cell pain in the ED, pain is assessed through patient's self-report supported by physical examination, lab results and other investigations if needed. ED physicians should take a detailed history to assess the pain, focusing on its triggering factors, site, severity, and if its nature is like previous episodes or not. The presence of an unusual feature of pain suggests that the pain could be due to causes other than VOC that the physician should assess. Physical examination should be directed at the pain sites, but the physician should also look for infection signs as pneumonia, osteomyelitis, or sepsis; splenomegaly; and priapism in males. The standard laboratory investigations include a complete blood count and reticulocyte count. Additional laboratory tests may be needed if fever or other signs are present for further assessment [15].

Severe acute SCD pain must be assessed and treated rapidly with the first dose of analgesic to be given within 15 - 30 minutes of presentation, which the American Pain Society; the National Institutes of Health; NHLBI, Division of Blood Diseases; and the Sickle Cell Society in the United Kingdom recommends [16,17]. This is advocated by the Cooperative Study of Sickle Cell Disease, in which 33% of the deaths were found to be during an acute VOC [18]. In our study, 56.4% of the ED physicians reported that the average time between presentation to the ED and administration of an analgesic is less than 30 minutes, while 37.6% stated that patients with SCD usually wait for 30-60 minutes before receiving any analgesic.

One of the most important factors that result in the delay in analgesic administration is the crowdedness of the EDs, which necessitates the ED physicians to focus on the life-threatening conditions and give them the priority to be treated before other patients with less urgent conditions. Emergency departments have complicated systems and limited supplies essential for critical care settings. There are multiple elements that make the ED management of patients with SCD to be inefficient; those involve insufficient resources, absence of management protocols, and biases which result in miscommunication, loss of trust, and worsening relationships between physicians and patients [5,6].

The most critical role for the Emergency departments is providing management of urgent cases. Otherwise, primary health care centers should provide management for stable cases that need regular care. A study found that patients with SCD who used to follow up with an outpatient health facility did not need to visit emergency departments as much as those who do not follow up regularly. This facility can also be utilized to manage SCD pan crisis which would decrease the load on the emergency departments and help patients with SCD to receive the appropriate management in a shorter duration [13].

As per NHLBI guidelines and Saudi MOH manual in Emergency Medicine, the first step in managing severe pain is hydration, which is done by 57.7% of the ED physicians included in our study. The precipitating factors and complications then should be assessed together with the pain severity to decide the type and dose of the analgesic required [7,19].

As recommended by NHLBI guidelines and Saudi MOH website, over the counter pain medications as Nonsteroidal anti-inflammatory drugs (NSAIDs) or acetaminophen can be used to subside mild-to-moderate pain, while persistent or severe pain should be managed with opioids [7,20]. Patients usually present to the ED after several trials of home medications to relieve their pain. When patients seek medical help, they are mostly in need of stronger or parenteral analgesics like morphine. To select the appropriate medication, the physician should take proper history focusing on the usual drugs and doses the patient uses, side effects, effective home medications, and the medications taken for the current pain. The physician should start the patient on the same dose of opioids that effectively subsided the pain in the previous visit for patients with recurrent pain episodes. Chronic use of opioids should be first excluded as it may result in the development of tolerance in which the patient needs a different drug or a higher dose of the same drug for the pain to be relieved [7].

In our study, most of the physicians (81.7%) whom hospitals that conduct lectures about SCD start with good hydration in managing VOC, while 57.3% of ED physicians working at hospitals that do not provide teaching sessions start the management with analgesics. Regarding the type of analgesic, both groups mostly (79.9%) start with weak analgesic then order stronger drug if the pain didn't improve.

We found that 65.8% of ED physicians usually choose morphine as the analgesic of choice for acute SCD painful episodes, with tolerance to be the most common side effect of analgesics seen in patients with SCD as reported by 88.6% of ED physicians. Chronic opioid use can result in physical dependence, which doesn't require treatment except when associated with withdrawal symptoms such as nausea, vomiting, diarrhea, dysphoria, nasal congestion, sweating, and seizures. The risk for developing withdrawal symptoms increases when opioids are used for longer than 5 - 7 days without tapering the doses.

Addiction is a psychological dependence that is related to social, psychological, and genetic factors. SCD patients are not considered addicts when they need opioids to relieve their acute pain. Recurrent ED visits by patients with SCD complaining of pain suggest that an

underlying undiagnosed etiology exists as avascular necrosis, bone infection or infarction, or that the pain management they receive is inadequate. This is known as pseudo addiction, in which patients seek out analgesics due to the presence of real pain that is usually not effectively treated, and it must be differentiated from addiction [7].

59.1% of physicians in our study reported that they participate in educating the patients about the importance of the compliance on long-term treatment and follow-up sessions with a hematology clinic. Physicians should educate the patients and their families about pain management, lifestyle modifications, and the advantage of using Hydroxyurea, which helps to reduce the frequency of pain attacks. Only 40.3% of the physicians in our study stated that their hospitals conduct educational sessions about SCD and its management. It's very important to educate ED nurses and physicians about pathophysiology, complications, and management of SCD crises especially VOC, to improve the care quality and thus reduce the chance to develop complications [6,7].

Conclusion

Sickle cell is considered as a globally widespread health issue with a significant impact on public health, with the VOC to be the most common cause of emergency department visits. In our study we found that most of the ED physicians depend on physical examination and vital signs respectively for the assessment of the pain in patients with SCD. 57.7% of the physicians stated that they start their management with good hydration, while 40.9% used to start with administering analgesia. Most of the physicians (56.4%) reported that the usual time gap between patients' presentation and analgesic administration is less than 30 minutes, while 37.6% stated that it usually takes 30 - 60 minutes. The most common analgesia type prescribed by most of ED physicians was found to be Morphine (65%). 132 physicians reported that tolerance is the most prevalent side effect of analgesics seen in SCD patients.

Recommendations

We recommend further research to identify sickle cell crisis management protocols in the emergency departments in Saudi Arabia and to assess the physicians' compliance with the protocols and its effects on patients' outcomes.

Conflict of Interest

There is no conflict of interest.

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