# Sickle Cell Disease: Core Concepts for the Emergency Physician and Nurse

## *Sickle Cell Pain*

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# Objectives

- Identify indicators of pain which are NOT reliable for individuals experiencing a vaso-occlusive Crisis (VOC)
- Accurately assign a triage category for an individual presenting with severe pain and a VOC

# Pretest- Question 1

The following are reliable, objective indicators of acute sickle cell disease (SCD) pain

a. Elevated blood pressure
b. Blood/Oxygen saturation level below 92%
c. Hemoglobin below 8
d. All of the above
e. None of the above

# Pretest- Question 2

An acute sickle-cell pain crisis with no other accompanying complications, & reported pain level of 8/10, is appropriately assigned a triage level of 3 using the Emergency Severity Index recommendations

a. True
b. False

# Epidemiology of SCD Pain

- Pain drives the majority of interactions with the healthcare system for patients with SCD
- Generally speaking, there are two types of SCD pain:
  - The vaso-occlusive crisis (VOC) – also known as a "sickle cell crisis"; an attack of acute pain due to vaso-occlusion that is the "hallmark symptom" of the disease
  - Chronic Pain : Due to accumulation of organ/tissue damage over time…may also be due to chronic levels of vaso-occlusion

# Epidemiology of SCD Pain

- Research has shown that the underlying burden of SCD pain is higher than most clinicians are aware
- In a study of approximately 31,000 SCD patient diary days, pain was noted on 54.5% of days[7]
- Unscheduled healthcare utilization, though, only occurred on 3.5% of days

# Epidemiology of SCD Pain[7]

- 55% of patients reported pain on at least 51% of their days
- 29% of patients reported pain on at least 96% of their days
- The “iceberg” model of SCD pain: a majority of the pain experienced by patients with SCD is not observed by healthcare providers

# Diagnosing Acute SCD Pain

- The following objective indicators can be reliably used in the diagnosis of acute SCD pain:
  - Lab work: None known
  - Radiographic findings: None known
  - Vital signs: None known
- As of this time, there are no objective indicators that can be used to reliably indicate the presence &/or severity of a VOC
- The patient's self-report is the gold-standard by which a VOC is identified

# Treating Acute SCD Pain

- Guidelines for the management of acute SCD pain in the ED typically promote the following principles:
  - Rapid clinical assessment
  - Involve the patient (i.e. ask about medicines/doses that typically work, what was taken at home & how much, how quality of current pain compares to typical acute pain episodes)
  - Caution: not all patients' require high doses of opioids.
  - Aggressive management…typically involving opioids. Opioids are required for many patients. Use doses that account for opioid tolerance developed from the patient's prior history with opioids.
  - Frequent re-assessment and re-administration of pain medicine if patient's pain not tolerable
  - Monitor for over-sedation

# Addiction & Substance-Abuse among Patients with SCD

- Clinician fears about contributing to, or causing, addiction to opioids among patients with SCD are a recognized barrier to the delivery of high quality pain management
- Multiple research studies have found the prevalence of substance abuse and addiction among patients with SCD to be lower than, or at most the same as, that found in the general population

# “High Utilizers” or “Frequent Flyers”

- There is a known subset of patients with SCD that contributes a disproportionate amount of ED utilization
- This high-utilizing subset also has been shown to have more severe disease requiring treatment

# Recommendations for Emergency Department Triage of SCD

- The Emergency Severity Index identifies acute sickle cell crisis as a condition that warrants a level 2 (high risk) emergency department triage assignment if pain report is ≥ 7/10:
  - "*Sickle cell disease requires immediate medical attention because of the severity of the patient's pain, which is caused by the sickle cells occluding small and sometimes large blood vessels. Rapid analgesic management will help prevent the crisis from progressing to the point where hospitalization will be unavoidable.*"[3]

# Patient Pain Coping Behaviors

- There is great variation in patient's ability to cope with pain and in the expressions they exhibit while in severe pain
- Patients may not express as much distress in their appearance as clinicians might assume given the patient's reported level of severe pain
- A patient with SCD is able to watch TV, talk on the telephone, talk with visitors, or appear relatively comfortable & still be in severe pain in need of aggressive treatment & close monitoring
- These behaviors & responses to pain are a subset of the coping skills patients have developed from years of experiencing painful episodes

# Clinical Scenario

- http://sickleemergency.duke.edu/content/crisis-experiences-people-sickle-cell-disease-seeking-health-care-pain
- Cut and paste this link into your browser to watch this short video

# Posttest- Question 1

The following are reliable, objective indicators of acute sickle cell disease (SCD) pain

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# Posttest- Question 2

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a. True
b. False

# Posttest Answers & Rationale

- ***Question 1***
  - Answer : e) None of the above
  - Rationale: There are no reliable, objective indicators of acute SCD pain. Patient self-report is the gold standard of pain measurement
- ***Question 2***
  - Answer: b) False
  - Rationale: The Emergency Severity Index recommendation for an acute SCD pain crisis with a report of severe (>7/10) pain is a level 2 triage rating (high-risk)

# References


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