Key Components of Pain Management for Children and Adults with Sickle Cell Disease

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KEYWORDS

• Sickle cell disease • Acute pain • Chronic pain syndrome • Opioids • Depression • Anxiety • Sleep

KEY POINTS

- The optimal management of acute pain episodes and chronic pain syndromes in sickle cell disease requires an understanding of the pharmacology principles of pain management.
- Pain management should be delivered using the biopsychosocial model, with interactions between biological, psychological, and social influences that contribute to pain addressed.
- Sickle cell disease pain management should target and keep individuals within the therapeutic window, maximizing analgesic effect and minimizing side effects.
- A complete sickle cell disease pain assessment should include screening for depression, anxiety, and sleep disturbances.

BACKGROUND OF INDIVIDUALS WITH SICKLE CELL DISEASE PAIN

Acute pain episodes are the most common complication of sickle cell disease (SCD), an inherited hemoglobinopathy affecting more than 3 million individuals worldwide.[1,2] Acute pain episodes are abrupt in onset, unpredictable, and account for the majority of health care use for SCD; however, these episodes are also frequently managed at home.[3] Acute pain episodes increase in frequency with age, and a chronic pain

Conflicts of Interest: The authors declare no competing financial interests.
Funding: National Institutes of Health, National Heart, Lung, and Blood Institute 1K23 HL114636-01A1 (A.M. Brandow).
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Hematol Oncol Clin N Am 32 (2018) 535–550
https://doi.org/10.1016/j.hoc.2018.01.014
hemonc.theclinics.com

syndrome evolves in 30% to 40% of adolescents and adults with SCD that significantly impairs functioning.[4,5]

Transition from Acute to Chronic Pain

The abrupt onset of acute pain episodes commonly occurs in the back, extremities, chest, and abdomen.[6] Temporally associated triggers for pain include, but are not limited to, acute infections, dehydration, asthma, cold temperatures, and the onset of menstruation; however, often no trigger is identified.[7–10] Acute pain episodes can start as early as in the first few months of life, increase in frequency with age, and can contribute to the development of a chronic pain syndrome.[3–5] The biologic basis for acute pain and the emergence of a chronic pain syndrome are likely different. Acute pain is caused by recurrent vasoocclusion from sickled erythrocytes with resultant ischemia–reperfusion injury, whereas chronic pain is likely driven by nervous system sensitization.[11] **Fig. 1** depicts the pain trajectory in individuals with SCD.

The diagnosis of a chronic pain syndrome in SCD is challenging, and includes many biologic, psychological, and sociologic risk factors. Traditionally, chronic pain is defined as pain persisting at least 3 to 6 months beyond the normal time for healing.[12] This definition often does not apply to individuals with SCD, because SCD pain develops over the lifetime. Evidence-based consensus diagnostic criteria, however, have been established for chronic pain syndrome in SCD.[13] A key component of these criteria includes: "Reports of ongoing pain on most days over the past 6 months either in a single location or multiple locations."[13]

Assessment of Individuals with Pain in Sickle Cell Disease

No objective measure can assess pain in children and adults with SCD. Thus, the cornerstone of pain management is trust between the affected individual in pain and the health care provider. Pain assessment must incorporate tools that account for the multidimensional aspects of pain. Classic pain assessments use unidimensional measures of pain intensity such as a numeric rating scale, the Wong-Baker Pain Scale, and the visual analog scale.[14,15] These scales are limited by the momentary assessment of pain and interindividual variability owing to differences in pain tolerance. Thus, the rating on a pain intensity scale should never be the sole determinant for the administration of analgesia.

Unfortunately, pain intensity scales do not assess the impact of pain on daily functioning, making them less useful for chronic pain. Instead, patient-reported outcome measures that capture multidimensional aspects of pain and the impact on functioning should be used. These tools include SCD-specific measures (PedsQL SCD Module, Adult Sickle Cell Quality of Life Measurement Information System)[16,17] and general measures (National Institutes of Health Patient-Reported Outcomes Measurement Information Systems).[18,19] Pain-specific tools with a 7- to 30-day recall period allow for assessment of pain over time and response to treatment.[16–19] Other multidimensional tools studied in SCD include, but are not limited to, the Youth Acute Pain Functional Ability Questionnaire,[20] Adolescent and Pediatric Pain Tool,[21] Brief Pain Inventory,[22] and McGill Pain Questionnaire.[23]

Assessment of SCD pain should elicit whether pain is acute, chronic, related to SCD, or all three. A clear discussion with the affected individual is required to distinguish between potential types of pain. Pain associated with "overuse syndrome," which is defined as pain from repetitive motions in daily activities, can be misunderstood and treated as acute SCD pain, chronic pain syndrome, or a prolonged acute pain episode. The temporal association of the new onset of pain