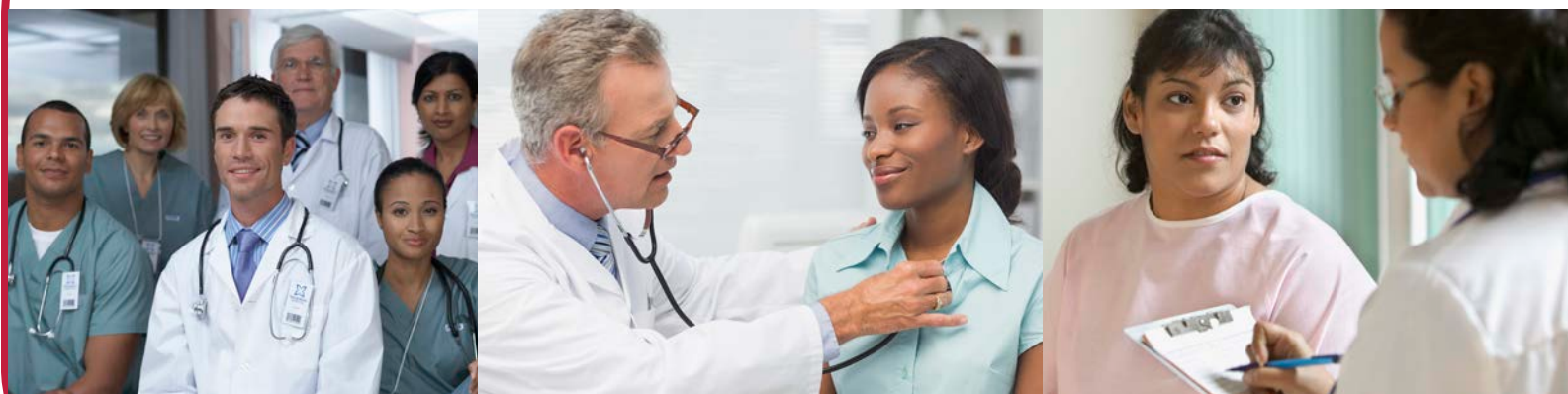


# Evidence-Based Management of Sickle Cell Disease

Expert Panel Report, 2014: Guide to Recommendations



**U.S. Department of Health and Human Services**  
National Institutes of Health  
National Heart, Lung, and Blood Institute

[www.nhlbi.nih.gov](http://www.nhlbi.nih.gov)

# Chapter 3: Managing Acute Complications of Sickle Cell Disease

## Vaso-Occlusive Crisis (VOC)

The recommendations labeled “consensus” in this section were based on recommendations developed by the American Pain Society (APS) or on panel expertise. The remaining recommendations are based on the evidence review conducted by the methodology team. These recommendations are intended to be for all settings where patients present with VOC.

1. In adults and children with SCD and pain,
  - When indicated, initiate diagnostic evaluation of causes of pain other than a VOC while beginning to treat pain.  
**(Consensus–Adapted)**
2. In adults and children with SCD and a VOC,
  - Determine characteristics, associated symptoms, location, and intensity of pain based on patient self-report and observation. If the VOC pain is atypical, investigate other possible etiologies of pain.  
**(Consensus–Adapted)**
  - Rapidly assess the patient’s recent analgesic use (opioid and nonopioid).  
**(Consensus–Adapted)**
  - Rapidly initiate analgesic therapy within 30 minutes of triage or within 60 minutes of registration.  
**(Consensus–Panel Expertise)**

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- Base analgesic selection on pain assessment, associated symptoms, outpatient analgesic use, patient knowledge of effective agents and doses, and past experience with side effects.

**(Consensus–Adapted)**

3. In adults and children with SCD and a VOC,
  - Use an individualized prescribing and monitoring protocol (written by the patient’s SCD provider) or an SCD-specific protocol whenever possible to promote rapid, effective, and safe analgesic management and resolution of the VOC.

**(Consensus–Panel Expertise)**

4. In adults and children with SCD and a VOC associated with mild to moderate pain who report relief with NSAIDs in the absence of contraindications to the use of NSAIDs, continue treatment with NSAIDs.

**(Moderate Recommendation, Low-Quality Evidence)**

5. In adults and children with SCD and a VOC associated with severe pain, rapidly initiate treatment with parenteral opioids.

**(Strong Recommendation, High-Quality Evidence)**

6. In adults and children with SCD and a VOC associated with severe pain,
  - Calculate the parenteral (IV or subcutaneous) opioid dose based on total daily short-acting opioid dose currently being taken at home to manage the VOC.

**(Consensus–Panel Expertise)**

- Administer parenteral opioids using the subcutaneous route when intravenous access is difficult.

**(Consensus–Panel Expertise)**

- Reassess pain and re-administer opioids if necessary for continued severe pain every 15–30 minutes until pain is under control per patient report.

**(Consensus–Adapted)**

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- Maintain or consider escalation of the dose by 25 percent until pain is controlled.  
**(Consensus–Panel Expertise)**
  - Reassess after each dose for pain relief and side effects.  
**(Consensus–Panel Expertise)**
  - Initiate around-the-clock opioid administration by patient-controlled analgesia (PCA) or frequently scheduled doses versus “as requested” (PRN) administration.  
**(Moderate Recommendation, Low-Quality Evidence)**
7. If ordering around-the-clock, continuous infusion of opioids via the PCA, carefully consider whether there is a need to withhold long-acting oral opioids to prevent over-sedation.  
**(Consensus–Panel Expertise)**
- If demand dosing only is ordered via the PCA, continue use of long-acting oral opioids.  
**(Consensus–Panel Expertise)**
  - At discharge, evaluate inpatient analgesic requirements, wean parenteral opioids prior to conversion to oral opioids, and adjust home dose of long- and short-acting opioid prescriptions to prevent opioid withdrawal after discharge.  
**(Consensus–Panel Expertise)**
8. In adults and children with SCD and a VOC, do not use meperidine unless it is the only effective opioid for an individual patient.  
**(Consensus–Adapted)**
9. In adults and children with a VOC, administer oral NSAIDS as an adjuvant analgesic in the absence of contraindications.  
**(Consensus–Adapted)**
10. In adults and children with a VOC who require antihistamines for itching secondary to opioid administration, prescribe agents orally, and do not re-administer with each dose of opioid in the acute VOC management phase. Re-administer every 4 to 6 hours if needed.  
**(Consensus–Panel Expertise)**

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11. To reduce the risk of acute chest syndrome in adults and children hospitalized for a VOC,
  - Encourage use of incentive spirometry while awake.  
**(Strong Recommendation, Moderate-Quality Evidence)**
  - Encourage ambulation and activity as soon as possible.  
**(Consensus–Panel Expertise)**
12. In adults and children with VOC, use adjunctive nonpharmacologic approaches to treat pain such as local heat application and distraction.  
**(Consensus–Adapted)**
13. In euvolemic adults and children with SCD and a VOC who are unable to drink fluids, provide intravenous hydration at no more than maintenance rate to avoid over-hydration.  
**(Consensus–Adapted)**
14. In adults and children with SCD and a VOC being treated with opioids, monitor for excessive sedation by measuring sedation with an objective measurement sedation scale and oxygenation levels.  
**(Consensus–Panel Expertise)**
15. Gradually titrate down parenteral opioids as VOC resolves.  
**(Consensus–Panel Expertise)**
16. In adults and children with SCD and a VOC, do not administer a blood transfusion unless there are other indications for transfusion (see the chapter “Blood Transfusion in the Management of Sickle Cell Disease” in this report).  
**(Moderate Recommendation, Low-Quality Evidence)**
17. In adults and children with SCD and a VOC with an oxygen saturation <95 percent on room air, administer oxygen.  
**(Consensus–Panel Expertise)**