



Complete Summary

GUIDELINE TITLE

Guideline for the management of acute and chronic pain in sickle cell disease.

BIBLIOGRAPHIC SOURCE(S)

Guideline for the management of acute and chronic pain in sickle cell disease. Glenview (IL): American Pain Society (APS); 1999 Aug. 98 p. (Clinical practice guideline; no. 1). [98 references]

COMPLETE SUMMARY CONTENT

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SCOPE

DISEASE/CONDITION(S)

Sickle-cell Disease (SCD)

GUIDELINE CATEGORY

Evaluation
Management
Treatment

CLINICAL SPECIALTY

Anesthesiology
Emergency Medicine
Family Practice
Hematology
Internal Medicine
Nursing
Pharmacology
Psychiatry

INTENDED USERS

Advanced Practice Nurses
Allied Health Personnel
Nurses
Pharmacists
Physician Assistants
Physicians

GUIDELINE OBJECTIVE(S)

- To aid physicians, nurses, and other healthcare professionals in managing acute and chronic pain associated with sickle-cell disease (SCD).
- To provide information for pain specialists unfamiliar with the dynamics of sickle-cell pain.

TARGET POPULATION

Patients with sickle-cell disease

INTERVENTIONS AND PRACTICES CONSIDERED

Assessment

1. Rapid assessment during a painful episode
 - Characteristics, location, and intensity (via self-report and/or observation)
2. Comprehensive assessment
 - Treatment history
 - Physical factors
 - Demographic and psychosocial factors
 - Dimensions of pain
 - Impact of pain on functioning
3. Pain assessment instruments
 - Attia Behavioral Pain Scale
 - Children's Hospital of Eastern Ontario (CHEOPS)
 - Oucher
 - PokerChip Tool
 - Wong-Baker Faces
 - Word Graphic Rating Scale
 - Numeric Pain Intensity Scale
 - Visual Analog Scale
 - Adolescent Pediatric Pain Tool (APPT)
 - Brief Pain Inventory (BPI)
 - Brief Pain Inventory Short Form
 - Memorial Assessment Card (MPAC)
 - McGill Pain Questionnaire
 - Pain Diary
 - Multidimensional Pain Inventory

Treatment

1. Pharmacologic treatments including:
 - Acetaminophen and over-the-counter nonsteroidal anti-inflammatory drugs (NSAIDs)
 - Prescription nonsteroidal anti-inflammatory drugs
 - Parenteral nonsteroidal anti-inflammatory drugs
 - Parenteral or oral steroids
 - Opioids and combination opioid/nonsteroidal anti-inflammatory drug preparations
 - Adjuvant medications to increase analgesic effect of opioids, reduce side effects of primary analgesic medications, or to manage other symptoms associated with sickle-cell disease
2. Selection of the appropriate opioid starting dose
3. Calculation of rescue or supplemental opioid dose
4. Weaning or tapering
5. Patient-controlled analgesia
6. Behavioral interventions including:
 - Relaxation
 - Deep breathing
 - Behavior modifications
 - Biofeedback
 - Exercise
 - Self-hypnosis
7. Physical interventions including:
 - Hydration
 - Heat
 - Massage
 - Hydrotherapy
 - Ultrasound
 - Acupuncture/acupressure
 - Transcutaneous electrical nerve stimulation (TENS)
 - Physical therapy
8. Psychological interventions including:
 - Education
 - Cognitive therapies
 - Hypnotherapy
 - Imagery
 - Distraction
 - Social support
 - Psychotherapy

MAJOR OUTCOMES CONSIDERED

- Patient-reported pain intensity recorded with standard pain scales
- Patient satisfaction with pain relief

METHODOLOGY

METHODS USED TO COLLECT/SELECT EVIDENCE

Searches of Electronic Databases

DESCRIPTION OF METHODS USED TO COLLECT/SELECT THE EVIDENCE

Literature from January 1965 through January 1999 was searched using the MEDLINE database, which produced 487 citations containing the keywords pain and sickle cell anemia. The search was duplicated using the National Library of Medicine at the National Institute of Health's (NIH's) PubMed and Grateful Med, with similar results. The abstracts were searched to identify articles reporting on research. Case reports, letters to the editor, articles describing diagnostic techniques, and surveys reporting the incidence of sickle-cell disease were excluded. One hundred and twenty-four articles were reviewed and rated for scientific rigor. Forty-one studies that described pain measurement instruments used with sickle-cell pain populations, or studies that used experimental or quasi-experimental designs to test relationships between specific interventions and pain were identified.

NUMBER OF SOURCE DOCUMENTS

Forty-one studies

METHODS USED TO ASSESS THE QUALITY AND STRENGTH OF THE EVIDENCE

Expert Consensus (Committee)
Weighting According to a Rating Scheme (Scheme Given)

RATING SCHEME FOR THE STRENGTH OF THE EVIDENCE

Type of Evidence

- I. Meta-analysis of multiple well-designed controlled studies.
- II. At least one well-designed experimental study.
- III. Well-designed, quasi-experimental studies, such as nonrandomized controlled, single-group pre-post, cohort, time series, or matched-case controlled studies.
- IV. Well-designed nonexperimental studies, such as comparative and correlational descriptive and case studies.
- V. Case reports and clinical examples.

METHODS USED TO ANALYZE THE EVIDENCE

Review of Published Meta-Analyses
Systematic Review with Evidence Tables

DESCRIPTION OF THE METHODS USED TO ANALYZE THE EVIDENCE

Not applicable

METHODS USED TO FORMULATE THE RECOMMENDATIONS

Expert Consensus

DESCRIPTION OF METHODS USED TO FORMULATE THE RECOMMENDATIONS

Not stated

RATING SCHEME FOR THE STRENGTH OF THE RECOMMENDATIONS

Strength and Consistency of Evidence Supporting the Recommendations

- A. There is evidence of type I or consistent findings from multiple studies of types II, III, or IV.
- B. There is evidence of types II, III, or IV, and findings are generally consistent.
- C. There is evidence of types II, III, or IV, but findings are inconsistent.
- D. There is little or no evidence, or there is type V evidence only.

Panel Consensus: Practice recommended based on the opinions of experts in pain management.

COST ANALYSIS

A formal cost analysis was not performed and published cost analyses were not reviewed.

METHOD OF GUIDELINE VALIDATION

External Peer Review
Internal Peer Review

DESCRIPTION OF METHOD OF GUIDELINE VALIDATION

A draft of this guideline underwent peer review by 61 experts in the management of sickle-cell pain and was revised based on their critiques and suggestions. With the cooperation of the Sickle Cell Disease Association of America, the revised draft was reviewed by a subset of the peer reviewers and 12 clinicians in pilot sites, who evaluated it for clarity and usefulness in practice (peer reviewers, site reviewers, and technical consultants are listed in Appendix D of the original guideline document). After further modification, the guideline was submitted to the American Pain Society Board of Directors, who approved it for dissemination.

RECOMMENDATIONS

MAJOR RECOMMENDATIONS

The following provides a summary of the recommendations presented in the guideline document. The reader is directed to the original guideline for a detailed discussion of each of the following topics.

The type of evidence (I-V) and the strength and consistency of evidence grades (A-D) are defined after the Major Recommendations. Citations in support of individual recommendations are identified in the original guideline document.

Pain Assessment in Sickle-Cell Disease

1. Clinicians should ask about pain, and patients' self-reports should be the primary source of assessment, with the exception of infants, for whom behavioral observations are the primary source of assessment.
Grade of evidence for recommendation: B
2. For rapid assessment of pain during an acute painful event, clinicians should select a simple measurement of pain intensity, reassess frequently, and record the measurement for treatment evaluation.
Grade of evidence for recommendation: B
3. A comprehensive biopsychosocial clinical assessment should be done yearly, and more often for patients with frequent pain.
Grade of evidence for recommendation: Panel consensus
4. To promote adequate pain management, patients should be reassessed frequently and asked how much their pain has been relieved after the first treatment and after subsequent treatment adjustments.
Grade of evidence for recommendation: Panel consensus
5. When clinicians consistently observe a disparity between patients' verbal self-report of their pain and their ability to function, further assessment should be performed to ascertain the reason for disparity.
Grade of evidence for recommendation: Panel consensus
6. Clinicians should understand and describe the pain in sufficient detail so that therapy can be tailored to the individual needs of patients.
Grade of evidence for recommendation: Panel consensus
7. Developmental state, chronological age, functional status, cognitive abilities, and emotional status should be considered in the choice of assessment methods and tools.
Grade of evidence for recommendation: B

Treatment of Pain in Sickle-Cell Disease

8. Pain management should be aggressive to ease the pain and enable patients to attain maximum functional ability.
Grade of evidence for recommendation: Panel consensus
9. Analgesics are the foundation for the management of sickle pain; their use should be tailored to the individual patient.
Grade of evidence for recommendation: B
10. Pharmacological management of mild-to-moderate-pain should include nonsteroidal anti-inflammatory drugs (NSAIDs) or acetaminophen, unless there is a contraindication. When mild-to-moderate pain persists, an opioid should be added. Treatment of persistent or moderate-to-severe pain should be based on increasing the opioid strength or dose.
Grade of evidence for recommendation: A
11. The type of oral opioid preparation used should be based on the characteristics and expected duration of the pain. If the patient's pain typically is of short duration (less than 24 hours), opioids or opioid formulations with short duration of action are appropriate, and they have the additional advantage of quicker onset of action. For patients whose pain requires several or many days to resolve, a sustained-release opioid preparation is more convenient and provides a more consistent analgesia.
Grade of evidence for recommendation: Panel consensus

12. Meperidine should not be used if frequent large doses or long treatment durations are anticipated.
Grade of evidence for recommendation: B
13. Opioid tolerance and physical dependence are expected with long-term opioid treatment and should not be confused with psychological dependence (addiction). Addiction is a pattern of compulsive drug use characterized by a continued craving for an opioid and a need to use the opioid for effects other than pain relief.
Grade of evidence for recommendation: B
14. Sedatives and anxiolytics alone should not be used to manage pain, because they can mask the behavioral response to pain without providing analgesia.
Grade of evidence for recommendation: B
15. Severe pain should be considered a medical emergency, and timely and aggressive management should be provided until the pain becomes tolerable.
Grade of evidence for recommendation: Panel consensus
16. Equianalgesic doses of oral opioids should be prescribed for home use when necessary to maintain the relief achieved in the emergency department or day hospital, or for a recurrence of severe pain.
Grade of evidence for recommendation: Panel consensus
17. Care should be taken to appropriately taper opioids in patients at risk for withdrawal symptoms.
Grade of evidence for recommendation: B

Psychological, Behavioral, and Physical Interventions

18. Because of changes in the disease and related pain, as well as changes in the patient's cognitive and psychological status, information about the pain should be communicated to the patient and family as an integral part of ongoing treatment.
Grade of evidence for recommendation: B
19. Analgesic medications are the mainstay of sickle-pain management, but they should be used with psychological, behavioral, and physical modalities.
Grade of evidence for recommendation: Panel consensus
20. Within a developmental framework, assessment and treatment of pain should be optimized early to provide a foundation for building further constructive pain management interventions as individuals proceed through life.
Grade of evidence for recommendation: B
21. Cognitive therapies should be used to enhance active coping strategies and reduce negative thinking.
Grade of evidence for recommendation: B

Definitions:

Type of Evidence

- I. Meta-analysis of multiple well-designed controlled studies.
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- IV. Well-designed nonexperimental studies, such as comparative and correlational descriptive and case studies.

V. Case reports and clinical examples.

Strength and Consistency of Evidence

- A. There is evidence of type I or consistent findings from multiple studies of types II, III, or IV.
- B. There is evidence of types II, III, or IV, and findings are generally consistent.
- C. There is evidence of types II, III, or IV, but findings are inconsistent.
- D. There is little or no evidence, or there is type V evidence only.
Panel Consensus: Practice recommended based on the opinions of experts in pain management.

CLINICAL ALGORITHM(S)

Clinical algorithms for pain assessment, treatment, and intravenous titration approaches are provided.

EVIDENCE SUPPORTING THE RECOMMENDATIONS

TYPE OF EVIDENCE SUPPORTING THE RECOMMENDATIONS

- The recommendations are based on an extensive review and analysis of the available scientific literature. When the research clearly supported a particular course of action, the studies were used to formulate a recommendation. When the findings of the studies conflicted or were equivocal, or when there were no systematic studies related to an aspect of management, research from related patient populations and the consensus of experts were used to make recommendations. (Sources of expert consensus included guidelines published by the New England Regional Genetics Group [Shapiro BS, Schechter NL, Ohene-Frempong K. Sickle-cell disease related pain. Quick reference guide for clinicians: assessment and management. Mt. Desert (ME): New England Regional Genetics Group, 1994] and the publication "Management and Therapy of Sickle-Cell Disease" [Reid CC, Charache S, Lubin B, Johnson C, Ohene-Frempong K (eds). Management and therapy of sickle-cell disease (3rd ed.). Bethesda (MD): National Heart, Lung and Blood Institute, 1995 (Publication No. 96-2117)]).
- The type of supporting evidence is identified for each recommendation (see "Major Recommendations").

BENEFITS/HARMS OF IMPLEMENTING THE GUIDELINE RECOMMENDATIONS

POTENTIAL BENEFITS

- Improvement in the management of pain for patients with sickle-cell disease.
- More comfort and improved quality of life for patients with sickle-cell disease.
- Improvement in professional practice and advance of pain-related research and education.

POTENTIAL HARMS

- Nonsteroidal anti-inflammatory drugs are not benign. Because patients with sickle cell disease have varying degrees of hepatic impairment, acetaminophen may be contraindicated in the presence of hepatic failure. All nonsteroidal anti-inflammatory drugs can be associated with renal failure, especially when used on a long-term basis. In addition, mild bleeding resulting from nonsteroidal anti-inflammatory drug-induced gastritis can be clinically dangerous for patients with sickle cell disease.
- Parenteral and oral steroids pose significant risks of gastritis, peptic ulcers, and osteoarthritis, as well as acceleration of avascular necrosis. There are also concerns about immunosuppression resulting from prolonged or frequent usage.
- Hypoventilation and respiratory depression can occur with the use of fentanyl.
- Common side effects of opioids include, sedation, nausea, and pruritis. Constipation and dysphoria occur on occasion. Respiratory depression (respiratory rate less than 10 per minute) is a rare side effect of opioid use.
- Opioid tolerance and physical dependence are expected pharmacological properties associated with long-term opioid use.

Subgroups Most Likely to be Harmed:

- Patients with hepatic impairment.
- Patients with gastritis, peptic ulcers, coagulopathies, and renal failure.
- Patients who require long-term use of opioids.

IMPLEMENTATION OF THE GUIDELINE

DESCRIPTION OF IMPLEMENTATION STRATEGY

An implementation strategy was not provided.

INSTITUTE OF MEDICINE (IOM) NATIONAL HEALTHCARE QUALITY REPORT CATEGORIES

IOM CARE NEED

Getting Better
Living with Illness

IOM DOMAIN

Effectiveness
Patient-centeredness

IDENTIFYING INFORMATION AND AVAILABILITY

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ADAPTATION

Not applicable: The guideline was not adapted from another source.

DATE RELEASED

1999 Aug

GUIDELINE DEVELOPER(S)

American Pain Society - Professional Association

SOURCE(S) OF FUNDING

This guideline was developed under funding from the American Pain Society (APS).

A list of corporate donors to the American Pain Society Guidelines Program is available on request from the American Pain Society.

GUIDELINE COMMITTEE

1999 Sickle-Cell Pain Guideline Panel

COMPOSITION OF GROUP THAT AUTHORED THE GUIDELINE

Panel Members: Lennette J. Benjamin, MD; Carlton D. Dampier, MD; Ada Jacox, PhD RN; Victoria Odesina, MS RN; David Phoenix, PhD; Barbara S. Shapiro, MD; Maureen Strafford, MD; Marsha Treadwell, PhD

FINANCIAL DISCLOSURES/CONFLICTS OF INTEREST

All members of the APS's Sickle-Cell Pain Management Guideline Panel have submitted a Conflict of Interest Disclosure Form, which has been reviewed by the APS Executive Director, who has determined that no conflict of interest exists with any individual panel member.

GUIDELINE STATUS

This is the current release of the guideline.

An update is not in progress at this time.

GUIDELINE AVAILABILITY

Electronic copies: Not available at this time.

Print copies: Available for purchase (\$15 nonmembers; \$10 members) from the American Pain Society (APS), 4700 W. Lake Avenue, Glenview, IL 60025-1485; Web site, www.ampainsoc.org. Orders can be placed via telephone, (847) 375-

4715 or by fax, (847) 375-4777. An [information request form](#) is also available at the Society's Web site.

AVAILABILITY OF COMPANION DOCUMENTS

The following is available:

- Guideline for the management of acute pain in sickle-cell disease. Quick reference guide for emergency department clinicians. Glenview (IL): American Pain Society, 2001 Apr.

Electronic copies: Not available at this time.

Print copies: Available for purchase from the American Pain Society (APS), 4700 W. Lake Avenue, Glenview, IL 60025-1485; Web site, www.ampainsoc.org. Orders can be placed via telephone, (847) 375-4715 or by fax, (847) 375-4777. An [information request form](#) is also available at the Society's Web site.

PATIENT RESOURCES

None available

NGC STATUS

This summary was completed by ECRI on March 15, 2001. The information was verified by the guideline developer as of April 3, 2001.

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