



Nursing Practice Guidelines: Care of the Patient with Sickle Cell Disease Experiencing Pain

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Sickle Cell Nurses and Physician Assistants

Introduction

The International Association of Sickle Cell Nurses and Physician Assistants has developed this guideline with the intention of promoting interaction among disciplines to ensure consistency of practice for patients with sickle cell disease experiencing pain. This nursing practice guideline applies to inpatient or outpatient management of acute or chronic pain. This nursing practice guideline is not intended to limit or dictate pain management practices.

Pain, the hallmark clinical manifestation of sickle cell disease, can occur as early as 4-6 months of age and is the leading reason for emergency department visits and hospitalizations. Pain from sickle cell disease, a result of vaso-occlusion, is unique in its pathophysiology and presentation. Vaso-occlusion is caused by sickled cells occluding capillaries and small blood vessels. This obstruction of blood flow causes tissue hypoxia and ischemia. Hypoxia in turn further increases sickling leading to a cyclic worsening of the painful event. Pain is subjective and can occur anywhere in the body. Sickle cell pain may be acute, chronic or a combination of both.

Assessment

- Onset
- Duration
- Location
- Quality
 - Typical or atypical
 - Impact on daily functioning
- Intensity
 - JACHO guidelines
 - Institutional policy
 - Pain assessment tools
- Prior interventions
- Hydration status
- Usual medications, dosage and side effects, medication allergies
- Usual non-pharmacological interventions



- Level of consciousness, developmental level and cognitive function
- Cultural influences
- Psychosocial support
- Usual coping mechanisms

Pain may have non-sickle cell etiology: menstrual cramps, trauma, appendicitis, pulmonary embolism/infarction, constipation and any other co-morbidities that may cause pain.

Intervention

The goal is prompt, effective pain management to the satisfaction of the patient. Because pain is a cumulative process, the window for opportunity to gain effective control is limited.

- Administer pain medications as soon as possible, ideally within 30 minutes of presentation.
 - Opioids
 - Patient controlled analgesia pump (PCA)
 - Scheduled dosing
 - NSAIDS
 - Adjuvants: sleep aids, anxiolytics and others as indicated
 - Continue home medications as prescribed
- Administer prophylaxis for constipation,
- Administer medications to treat pruritis, nausea, and over sedation as needed
- Incentive spirometer, ambulation
- Facilitate non-pharmacologic interventions: distraction, relaxation techniques, heat, massage, TENS units, biofeedback,
- Ensure adequate hydration (PO, IV)
- Reassess pain level at least hourly
- Vital signs, level of consciousness/sedation, pulse oximeter hourly
- Consultation with other services as indicated
 - Social work
 - Pastoral Care
 - Child life
 - Respiratory therapy
 - PT/OT
 - Mental health services
 - Patient Representatives/Advocates
 - Pain Service
 - Community support services

Education

- Adequate hydration
- Encourage adherence to prescribed home medications (hydroxyurea, other red cell modifiers, regularly scheduled anti inflammatories and/or pain control meds)
- Routine medical follow-up

- Encourage follow up with specialty referrals
- Support group participation
- Address barriers to health care maintenance: cultural influences, lack of insurance, financial stressors, geographic barriers, mental health factors
- Appropriate administration of home pain medications:
 - Dosage
 - Dosing schedule
 - When to begin and end
 - Maintain timely refills
- Side effects of pain medications
- Discuss tolerance vs addiction to pain meds
- Self- management strategies
 - Home regimen
 - Relaxation techniques
 - Massage
 - Bio feedback
 - Guided Imagery
- Nutrition and hydration
- Avoid known pain triggers
- Signs/symptoms to report to Hematologist
 - When to go to Ed/Treatment Center

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