

Guideline for the Management of Acute Pain in Sickle-Cell Disease in the Emergency Department

This guide was prepared to provide emergency department clinicians with quick access to guidelines for the management of acute pain in sickle-cell disease. Severe pain should be considered a medical emergency that needs timely and aggressive treatment to the best relief possible. These recommendations should be used in conjunction with *Guidelines for the Management of Acute and Chronic Pain in Sickle-Cell Disease* (American Pain Society, 1999).

Common Pain States Associated with Sickle-Cell Disease			
Pain States	Clinical Signs and Symptoms	Signs/Underlying Cause	Special Features/Considerations
• Acute painful event	<ul style="list-style-type: none"> • Sudden onset • Pain in any and all parts of body 	<ul style="list-style-type: none"> • Vaso-occlusion • Endothelial damage • Inflammation 	<ul style="list-style-type: none"> • Unpredictable, recurrent • Great variability • All ages
• Acute hand-foot syndrome (dactylitis)	<ul style="list-style-type: none"> • Painful dorsal swelling of hands and feet 	<ul style="list-style-type: none"> • Symmetrical infarcts of metacarpal and metatarsal bones due to obstruction of developing blood vessels 	<ul style="list-style-type: none"> • More common in childhood • Often first manifestation of disease (occurring as early as 6 months of age)
• Acute inflammation of joints	<ul style="list-style-type: none"> • Painful swollen joints 	<ul style="list-style-type: none"> • Vaso-occlusion/injury • Inflammation • Infected joints • Gout 	<ul style="list-style-type: none"> • May accompany dactylitis • Acute flare-ups as isolated events • Septic arthritis is rare but may occur
• Acute chest syndrome	<ul style="list-style-type: none"> • Chest pain, particularly rib and substernal area • Chest pain posteriorly (upper back) • Fever, tachypnea, and/or hypoxia 	<ul style="list-style-type: none"> • Pulmonary infiltrates • May be associated with infarction or infection • Unilateral pain (splinting from atelectasis) 	<ul style="list-style-type: none"> • May require exchange transfusion and can be fatal • Common cause of mortality in children and adults
• Splenic sequestration	<ul style="list-style-type: none"> • Left upper-quadrant pain • Marked pallor • Sudden decrease in hemoglobin concentration • Enlarged spleen 	<ul style="list-style-type: none"> • Blood trapped in the spleen 	<ul style="list-style-type: none"> • Can be catastrophic in children, with possibility of circulatory collapse • Insidious onset in adults • Occurs in older children and adults with HBSC and sickle beta thalassemia
• Intrahepatic sickling or hepatic sequestration	<ul style="list-style-type: none"> • Right upper-quadrant pain • Sudden decrease in hemoglobin • Enlarged liver 	<ul style="list-style-type: none"> • Blood pooling in the liver 	<ul style="list-style-type: none"> • Occurs more commonly in adults
• Abdominal and intra-abdominal pain	<ul style="list-style-type: none"> • Jaundice • Diffuse abdominal pain • Enlarged spleen 	<ul style="list-style-type: none"> • Cholelithiasis • Constipation secondary to opioid therapy • Splenic infarction 	<ul style="list-style-type: none"> • Can be initial manifestation of acute chest syndrome • Involve surgery if severe symptoms
• Priapism	<ul style="list-style-type: none"> • Painful erection 	<ul style="list-style-type: none"> • Sickling in sinusoids of penis 	<ul style="list-style-type: none"> • May last for a few hours (acute and brief) to days (acute and prolonged), or may be chronic or stuttering (intermittent)
• Avascular necrosis of femur or humerus	<ul style="list-style-type: none"> • Prolonged, constant bone pain • Shoulder pain • Knee pain • Hip pain 	<ul style="list-style-type: none"> • Associated with bone infarction, sickle arthritis 	<ul style="list-style-type: none"> • Physical therapy may be useful for reducing pain and maintaining function
• Chronic neuropathic pain	<ul style="list-style-type: none"> • Pain in back, lower extremities, other sites • Spontaneous • Lancing • Burning 	<ul style="list-style-type: none"> • Older adults: disc disease, infections • Collapsed vertebrae • Iron overload neuropathy 	<ul style="list-style-type: none"> • Must be considered in patients who are unresponsive to opioids • Treatment modalities may require days or weeks before taking effect • Creates chronic pain state