Management of painful vaso-occlusive crisis of sickle-cell anemia: consensus opinion.


Source

Pharmaceutical Research Institute, Albany College of Pharmacy and Health Sciences, Albany, New York 12144, USA. shaker.mosua@acphs.edu

Abstract

Sickle-cell disease (SCD) is a wide-spread inherited hemolytic anemia that is due to a point mutation, leading to the substitution of valine for glutamic acid, causing a spectrum of clinical manifestations in addition to hemolysis and anemia. Acute painful crisis is a common sequela that can cause significant morbidity and negatively impact the patient's quality of life. Remarkable improvements in the understanding of the pathogenesis of this clinical syndrome and the role of cell adhesion, inflammation, and coagulation in acute painful crisis have led to changes in the management of pain. Due to the endemic nature of SCD in various parts of the Middle East, a group of physicians and scientists from the United States and Middle East recently met to draw up a set of suggested guidelines for the management of acute painful crisis that are reflective of local and international experience. This review brings together a detailed etiology, the pathophysiology, and clinical presentation of SCD, including the differential diagnoses of pain associated with the disease, with evidence-based recommendations for pain management and the potential impact of low-molecular-weight heparin (LMWH), from the perspective of physicians and scientists with long-term experience in the management of a large number of patients with SCD.