Challenges to management of pain in sickle cell disease

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Abstract

Sickle Cell Disease (SCD) is one of the most common blood disorders in the world. Pain is the primary reason for which individuals with SCD interact with the healthcare system. Generally speaking, there are two types of SCD pain: vaso-occlusive pain (or sickle cell disease crisis) and chronic pain caused by an accumulation of organ and tissue damage over time. However, despite its frequency, we have limited understanding about what causes pain in sickle cell disease, how best to manage pain in SCD and (most importantly) how to prevent pain in SCD. For medical providers, pain is also an elusive target due to the difficulty in objectively measuring pain and the importance of relying on patient reported outcomes. To face the challenges in managing pain in SCD, we will review the current understanding of the pathophysiology of vaso-occlusion, the multiple dimensions of the pain experience, and the current methods of measuring and managing pain. We will also review new pharmacologic agents undergoing clinical trials in SCD that will help to prevent pain and improve outcomes in SCD.