

patients taking hydroxyurea alone revealed a 40% reduction in mortality and decreased frequency of VOC, ACS, hospital admissions, and need for transfusions, thus making SCD crises milder ([Anderson, 2006](#); [Strouse, Lanzkron, Beach, et al, 2008](#); [Voskaridou, Christoulas, Bilalis, et al, 2010](#)). Pediatric studies have shown that hydroxyurea can be safely used in children ([Wang, Ware, Miller, et al, 2011](#); [Zimmerman, Schultz, Davis, et al, 2004](#)).

Allogeneic hematopoietic stem cell transplantation (HSCT) offers a curative treatment for children with SCD with overall survival 92% to 95% and event-free survival of 82% to 86% ([Bernaudin, Socie, Kuentz, et al, 2007](#); [Haining, Duncan, and Lehmann, 2009](#); [Hsieh, Fitzhugh, Weitzel, et al, 2014](#); [Locatelli and Pagliara, 2012](#)).

Since SCD is an autosomal recessive disorder, curative strategies for correction, replacement, addition, or modulation of the globin gene continue to evolve in the basic and clinical research settings ([Meier and Miller, 2012](#)).

Quality Patient Outcomes: Sickle Cell Disease

- Early recognition of signs and symptoms of sickle cell anemia (SCA)
- Tissue deoxygenation minimized
- Sickle cell crisis prevented or quickly managed
- Pain appropriately managed
- Stroke prevented
- Prophylactic penicillin regimen followed
- Hypoxia prevented when surgery is necessary
- Pneumococcal, *H. influenzae* type b, and meningococcal vaccines administered

Nursing Care Management