

maximum of a 5-day course is commonly used to enhance the pain management effect. Patient-controlled analgesia (PCA) has been used successfully for sickle cell–related pain. PCA reinforces the patient's role and responsibility in managing the pain and provides flexibility in dealing with pain, which may vary in severity over time (see [Pain Management, Chapter 5](#)).

### **Drug Alert**

Meperidine (Demerol) is not recommended. Normeperidine, a metabolite of meperidine, is a central nervous system (CNS) stimulant that produces anxiety, tremors, myoclonus, and generalized seizures when it accumulates with repetitive dosing. Patients with SCD are particularly at risk for normeperidine-induced seizures ([Ellison, 2012](#); [Howard and Davies, 2007](#); [National Institutes of Health, National Heart, Lung, and Blood Institute, Division of Blood Disease and Resources, 2002](#)).

### **Prognosis**

The prognosis varies, but most patients live into the fifth decade. The greatest risk is usually in children younger than 5 years old, and the majority of deaths in these children are caused by overwhelming infection. Consequently, SCA is a chronic illness with a potentially terminal outcome. Physical and sexual maturation are delayed in adolescents with SCA. Although adults achieve normal height, weight, and sexual function, the delay may present problems to adolescents ([Heeney and Dover, 2009](#); [Redding-Lallinger and Knoll, 2006](#)).

Individuals with SCD who have higher levels of HbF tend to have a milder disease with fewer complications than those with lower levels ([Driscoll, 2007](#); [Meier and Miller, 2012](#)). Hydroxyurea is a US Food and Drug Administration–approved medication that increases the production of HbF, reduces endothelial adhesion of sickle cells, improves the sickle cell hydration and cell size, increases nitric oxide production (a vasodilator), and lowers leukocyte and reticulocyte counts ([McGann and Ware, 2011](#); [National Institutes of Health, National Heart, Lung, and Blood Institute, Division of Blood Disease and Resources, 2002](#); [Yawn, Buchanan, Afenyi-Annan, et al, 2014](#)). Long-term follow-up of