

Casella, and DeBaun, 2012; Yawn, Buchanan, Afenyi-Annan, et al, 2014). After a CVA, blood transfusions are usually given every 3 to 4 weeks to help prevent a repeat stroke. To reduce iron overload from chronic transfusion therapy, chelation therapy may be started (see later in chapter).

In children with recurrent life-threatening splenic sequestration, splenectomy may be a lifesaving measure. However, the spleen usually atrophies on its own through progressive fibrotic changes (**functional asplenia**) by 6 years of age in children with SCA. Prophylactic penicillin and pneumococcal vaccines have decreased the incidence of pneumococcal sepsis in children with SCD. Packed RBC transfusions are recommended not only for treatment of splenic sequestration but also stroke and used preoperatively accompanied with maintenance IV hydration for most surgical procedures in children with SCD.

VOC, the most common, severe, painful episode, is considered the clinical hallmark of SCD that is usually accompanied by increasing health care cost because of prolonged hospitalization (Ballas, 2011; McCavit, 2012; Raphael, Mei, Mueller, et al, 2012; Yawn, Buchanan, Afenyi-Annan, et al, 2014). The chronic nature of this pain can greatly affect the child's development. A multidisciplinary team (e.g., physician, psychologist, child life specialist, family, nurse, social worker) approach is best for vasoocclusive pain management that includes pharmacologic treatments, hydration, physical therapy, and non-pharmacologic and complementary treatment (e.g., prayer, spiritual healing, massage, heating pads, herbs, relaxation, breathing exercises, distraction, music, guided imagery, self-motivation, acupuncture, and biofeedback) (Ballas, 2011; Brandow, Weisman, and Panepinto, 2011; Meier and Miller, 2012; Redding-Lallinger and Knoll, 2006). When mild to moderate VOC is reported, nonsteroidal antiinflammatory medication (e.g., ibuprofen, ketorolac) or nonopioids (e.g., acetaminophen) are used initially. If these drugs are not effective alone, an opioid may be added. The dosages of both drugs are titrated (adjusted) to a therapeutic level. Opioids such as immediate- and sustained-release morphine, oxycodone, hydrocodone, hydromorphone (Dilaudid), and methadone are administered intravenously or orally for severe pain and are given around the clock. In conjunction with the opioid, IV ketorolac for a