

Zarkowsky HS, Gallagher D, Gill FM, et al. Bacteremia in sickle hemoglobinopathies. *J Pediatr*. 1986;109(4):579–585.

*Adapted from the Quality and Safety Education for Nurses website at <http://www.qsen.org>.

Oxygen therapy is of little therapeutic value unless the patient has hypoxia (Heeney and Dover, 2009). Severe hypoxia must be prevented because it causes massive systemic sickling that can be fatal. Oxygen administration is usually not effective in reversing sickling or reducing pain because the oxygen is unable to reach the enmeshed sickled erythrocytes in clogged vessels. In addition, prolonged administration of oxygen can depress bone marrow, further aggravating the anemia.

Another important component of care is the use of blood transfusions. Exchange RBC transfusion (erythrocytapheresis) is the replacement of sickle cells with normal RBCs. Exchange transfusion is a successful, rapid method of reducing the number of circulating sickle cells and therefore slowing down the vicious circle of hypoxia, thrombosis, tissue ischemia, and injury. Therapy including simple and exchange transfusions are used in life-threatening ACS and after acute overt stroke to prevent reoccurrence and further tissue damage (Velasquez, Mariscalco, Goldstein, et al, 2009; Meier and Miller, 2012; Wang and Dwan, 2013). A transcranial Doppler (TCD) test identifies the child with SCA or HgbS-B⁰ thalassemia who is at high risk for developing a CVA by monitoring the intracranial vascular flow (Driscoll, 2007; Kwiatkowski, Yim, Miller, et al, 2011; Meier and Miller, 2012). The TCD is performed yearly for children from 2 to 16 years old. The recommended treatment for children with confirmed abnormal TCD is chronic transfusion therapy (Armstrong-Wells, Grimes, Sidney, et al, 2009; Kwiatkowski, Yim, Miller, et al, 2011; Wang and Dwan, 2013). The duration of transfusion is indefinite, although current studies are addressing whether patients may be transitioned safely to hydroxyurea to prevent stroke (McCavit, 2012). Multiple transfusions carry the risk of transmission of viral infection, hyperviscosity, transfusion reactions, alloimmunization, and hemosiderosis (Driscoll, 2007; Heeney and Dover, 2009; Jordan,