Acute Chest Syndrome This pneumonia-like illness is the second most frequent acute sickle cell–related event. It occurs in >50% of patients, often more than once. Acute chest syndrome can be mild, especially in children, in whom it can result from viral infection, or devastating, where multiple lobes of the lung are affected with severe hypoxia, multiorgan failure, and death. Chest pain, cough, fever, and hypoxia and a pulmonary infiltrate on chest x-ray are the major diagnostic criteria. The etiology includes in situ thrombosis, emboli, any type of infection, and postoperative hypoventilation. Management in adults is dictated by the severity of the episode. Patients who are hypoxic and febrile are often admitted directly to the intensive care unit. Antibiotics are almost always used in febrile patients even though a causative bacterium is not often cultured. Supplemental O_2 is given for an O_2 saturation <95%. Overhydration and excessive opioids can compound dyspnea and hypoxia. Hypoxic patients who are febrile with leukocytosis and have more than a trivial infiltrate on x-ray are transfused. In the more severely ill patient, exchange transfusion is the preferred modality. When hemoglobin level or symptoms indicate the need for transfusion of the severely ill patient and hours are needed to arrange red cell exchange, simple or top-up transfusion should be started first. Simple transfusions also suffice for less severely affected patients. Most patients survive acute chest syndrome, but in the most severe cases, often caused by embolization of necrotic bone marrow, death can be rapid even with prompt and proper treatment. Thrombocytopenia, leukocyte counts in excess of 20,000/dL, and rapidly developing acute anemia often portend severe acute chest syndrome with the possibility of acute respiratory distress syndrome and multiorgan failure. Many adults have chronic lung disease that could be a sequela of acute chest syndrome, and asthma is very common in patients with sickle cell disease.

Osteonecrosis This painful and sometimes crippling complication that most often affects hips bilaterally occurs in about half of all patients with sickle cell anemia and is also common in HbSC disease; shoulders are less often affected. Beginning with chronic pain that can become severe, loss of function is often the final stage,