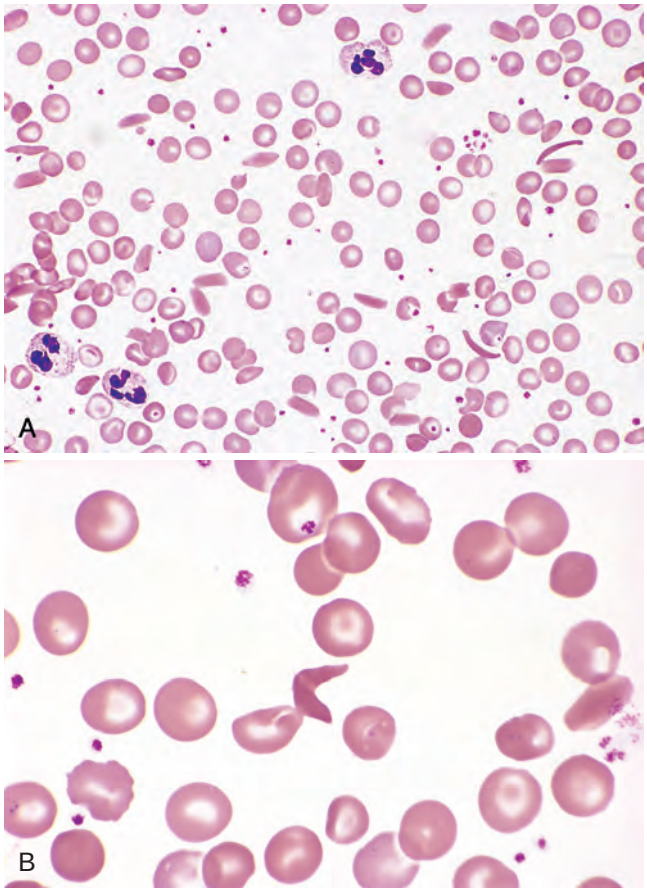

Figure 14-8 Sickle cell disease (peripheral blood smear). **A,** Low magnification shows sickle cells, anisocytosis, and poikilocytosis. **B,** Higher magnification shows an irreversibly sickled cell in the center. (Courtesy Dr. Robert W. McKenna, Department of Pathology, University of Texas Southwestern Medical School, Dallas, Texas.)

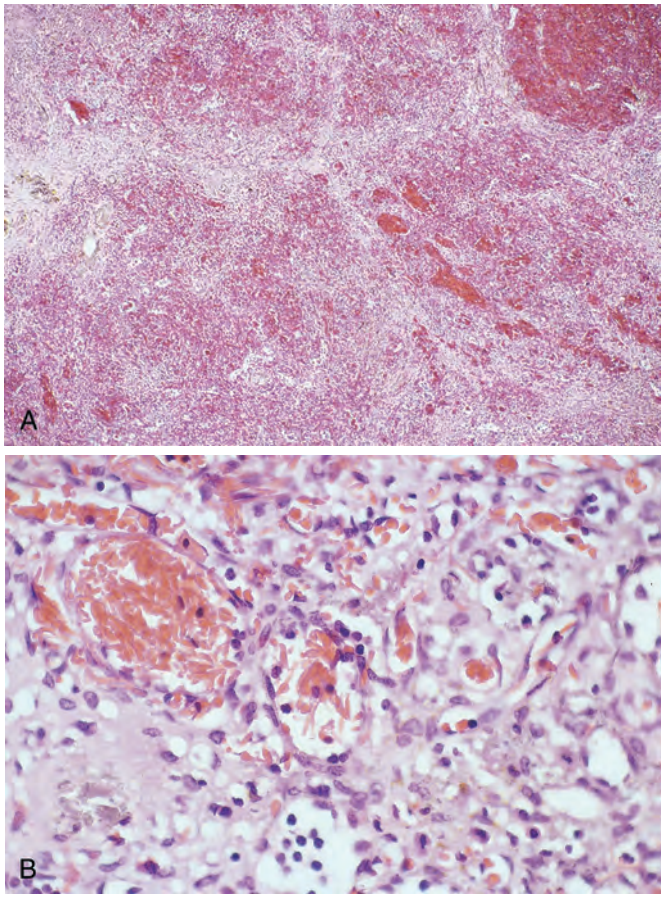

Figure 14-9 **A,** Spleen in sickle cell disease (low power). Red pulp cords and sinusoids are markedly congested; between the congested areas, pale areas of fibrosis resulting from ischemic damage are evident. **B,** Under high power, splenic sinusoids are dilated and filled with sickled red cells. (Courtesy Dr. Darren Wirthwein, Department of Pathology, University of Texas Southwestern Medical School, Dallas, Texas.)

fibrosis, and progressive shrinkage, so that by adolescence or early adulthood only a small nubbin of fibrous splenic tissue is left; this process is called **autosplenectomy** (Fig. 14-10). Infarctions caused by vascular occlusions can occur in many other tissues as well, including the bones, brain, kidney, liver, retina, and pulmonary vessels, the latter sometimes producing cor pulmonale. In adult patients, vascular stagnation in subcutaneous tissues often leads to leg ulcers; this complication is rare in children.

Clinical Features

Sickle cell disease causes a moderately severe hemolytic anemia (hematocrit 18% to 30%) that is associated with reticulocytosis, hyperbilirubinemia, and the presence of irreversibly sickled cells. Its course is punctuated by a variety of "crises." *Vaso-occlusive crises,* also called *pain crises,* are episodes of hypoxic injury and infarction that cause severe pain in the affected region. Although infection, dehydration, and acidosis (all of which favor sickling) can act as triggers, in most instances no predisposing cause is identified. The most commonly involved sites are the bones, lungs, liver, brain, spleen, and penis. In children, painful bone crises are extremely common and often difficult to distinguish from acute osteomyelitis. These frequently manifest as the *hand-foot syndrome* or dactylitis of the bones of the hands or feet, or both. *Acute chest syndrome* is a particularly dangerous type of vaso-occlusive crisis involving the lungs, which typically presents with fever, cough, chest pain, and pulmonary infiltrates. Pulmonary inflammation (such as may be induced by a

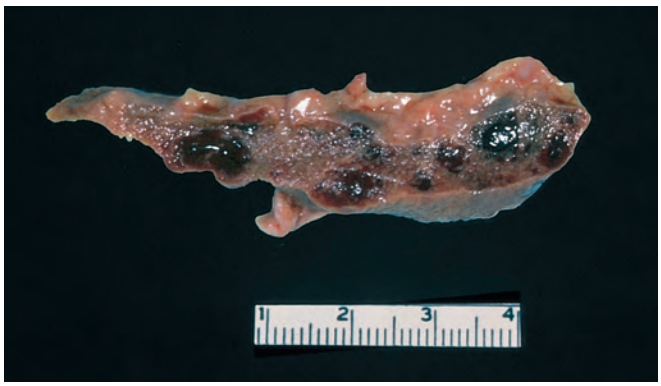

Figure 14-10 "Autoinfarcted" splenic remnant in sickle cell disease. (Courtesy Drs. Dennis Burns and Darren Wirthwein, Department of Pathology, University of Texas Southwestern Medical School, Dallas, Texas.)

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