FIGURE 98-4 Diagnosis of sickle cell disease. *A*. From *left to right*, high-performance liquid chromatography separation in sickle cell trait, sickle cell anemia, and HbSC disease. Beneath each chromatogram, the individual protein peaks are identified. ***B***. *Left:* Dense, elongated, and pointed cells are the irreversibly sickled cells characteristic of the sickle cell anemia and sickle cell-β^0 thalassemia. Target cells and nucleated red cells are also present. *Right:* Target cells, cells with squared ends of HbC crystals, cells folded like tacos, and contracted microspherocytes are typical of HbSC disease. *(Source: B [right]: Reproduced with permission from American Society of Hematology.)*

COMPLICATIONS

Complications of sickle cell disease can be grouped into those that likely are a consequence of sickle vasoocclusion and ones that appeared to be triggered by intravascular hemolysis. Although there is a relationship between these two limbs of pathophysiology, complications associated with vasoocclusion seem to respond best to induction of HbF. Some complications of disease are presented in **Table 98-3**. Early and effective treatment with hydroxyurea and the integration into management of new treatments discussed below should change this profile.

TABLE 98-3 Complications of Sickle Cell Disease