TABLE 79-2 DISEASES ASSOCIATED WITH SPLENOMEGALY GROUPED BY PATHOGENIC MECHANISM

Enlargement Due to Increased Demand for Splenic Function	
Reticuloendothelial system hyperplasia (for removal of defective erythrocytes)	Leishmaniasis
Spherocytosis	Trypanosomiasis
Early sickle cell anemia	Ehrlichiosis
Ovalocytosis	Disordered immunoregulation
Thalassemia major	Rheumatoid arthritis (Felty's syndrome)
Hemoglobinopathies	Systemic lupus erythematosus
Paroxysmal nocturnal hemoglobinuria	Collagen vascular diseases
Pernicious anemia	Serum sickness
Immune hyperplasia	Immune hemolytic anemias
Response to infection (viral, bacterial, fungal, parasitic)	Immune thrombocytopenias
Infectious mononucleosis	Immune neutropenias
AIDS	Drug reactions
Viral hepatitis	Angioimmunoblastic lymphadenopathy
Cytomegalovirus	Sarcoidosis
Subacute bacterial endocarditis	Thyrotoxicosis (benign lymphoid hypertrophy)
Bacterial septicemia	Interleukin 2 therapy
Congenital syphilis	Extramedullary hematopoiesis
Splenic abscess	Myelofibrosis
Tuberculosis	Marrow damage by toxins, radiation, strontium
Histoplasmosis	Marrow infiltration by tumors, leukemias, Gaucher's disease
Malaria	
Enlargement Due to Abnormal Splenic or Portal Blood Flow	
Cirrhosis	Splenic artery aneurysm
Hepatic vein obstruction	Hepatic schistosomiasis
Portal vein obstruction, intrahepatic or extrahepatic	Congestive heart failure
Cavernous transformation of the portal vein	Hepatic echinococcosis
Splenic vein obstruction	Portal hypertension (any cause including the above): "Banti's disease"
Infiltration of the Spleen	
Intracellular or extracellular depositions	Hodgkin's disease
Amyloidosis	Myeloproliferative syndromes (e.g., polycythemia vera, essential thrombocytosis)
Gaucher's disease	Angiosarcomas
Niemann-Pick disease	Metastatic tumors (melanoma is most common)
Tangier disease	Eosinophilic granuloma
Hurler's syndrome and other mucopolysaccharidoses	Histiocytosis X
Hyperlipidemias	Hamartomas
Benign and malignant cellular infiltrations	Hemangiomas, fibromas, lymphangiomas
Leukemias (acute, chronic, lymphoid, myeloid, monocytic)	Splenic cysts
Lymphomas	
Unknown Etiology	
Idiopathic splenomegaly	Iron-deficiency anemia
Berylliosis	

necessary for staging of patients with Hodgkin's disease only in those with clinical stage I or II disease in whom radiation therapy alone is contemplated as the treatment. Noninvasive staging of the spleen in Hodgkin's disease is not a sufficiently reliable basis for treatment decisions because one-third of normal-sized spleens will be involved with Hodgkin's disease and one-third of enlarged spleens will be tumor-free. The widespread use of systemic therapy to test all stages of Hodgkin's disease has made staging laparotomy with splenectomy unnecessary. Although splenectomy in chronic myeloid leukemia (CML) does not affect the natural history of disease, removal of the massive spleen usually makes patients significantly more comfortable and simplifies their management by significantly reducing transfusion requirements. The improvements in therapy of CML have reduced the need for splenectomy for symptom control. Splenectomy is an effective secondary or tertiary treatment for two chronic B cell leukemias, hairy cell leukemia and prolymphocytic leukemia, and for the very rare splenic mantle cell or marginal zone lymphoma. Splenectomy in these diseases may be associated with significant tumor regression in bone marrow and other sites of disease. Similar regressions of systemic disease have been noted after splenic irradiation in some types of lymphoid tumors, especially chronic lymphocytic leukemia and prolymphocytic leukemia. This has been termed the *abscopal effect*. Such systemic tumor responses to local therapy directed at the spleen suggest that some hormone or growth factor produced by the spleen may affect tumor cell proliferation, but this conjecture is not yet substantiated.

TABLE 79-3 DISEASES ASSOCIATED WITH MASSIVE SPLENOMEGALY[a]

Chronic myeloid leukemia	Gaucher's disease
Lymphomas	Chronic lymphocytic leukemia
Hairy cell leukemia	Sarcoidosis
Myelofibrosis with myeloid metaplasia	Autoimmune hemolytic anemia
Polycythemia vera	Diffuse splenic hemangiomatosis

[a]The spleen extends >8 cm below the left costal margin and/or weighs >1000 g.