Function of the Spleen: Cellular and humoral immunity (lymphoid), removal of old RBC’s, bacteria and other circulating particulates (monocyte-macrophage system), site of extramedullary hematopoiesis under certain circumstances (stem cells). Contains 25% of lymphoid mass of the body.

Splenomegaly: Usually >40% of normal size to be palpable, but not all palpable spleens will be abnormal, nor will all abnormal spleens been palpable (20% of spleens > 900 gms not palpable, nl = 150 gms)

->MNEMONIC FOR SPLENOMEGALY= “SPLEEN”

S – Sequestration: Congenital spherocytosis, congenital or acquired hemolytic anemias

P- Proliferation: Due to chronic inflammation or infection.
   -Inflammation: SLE, RA, Felty’s syndrome (RA, splenomegaly, neutropenia)
   -Infections. Viral: hepatitis, mono, CMV, Fungal
      -Bacterial: salmonella, brucella, TB
      -Parasitic: malaria, schistosomiasis, toxo, leishmaniasis, SBE

L- Liquid deposition disorders: Gaucher’s disease, Niemann- Pick, Amyloid, Glycogen storage disease

E- Endowment: As in congenital causes, ex: splenic hemangioma, hamartoma, or cysts

E- Engorgement: Splenic trauma with intracapsular hematoma, sequestration crisis in sickle cell, CHF, portal HTN from any cause

N -Invasion: Granulomatous or malignant disease. Usually indolent form of lymphoma, both acute and chronic leukemias, PCV, myeloid metaplasia, mets

Massive splenomegaly defined as splenomegaly that reaches to the pelvis or which has crossed the midline into the right lower or right upper abdominal quadrants. Narrowed differential: (NEJM 345: 682-687, 2001, NEJM 330:775-781, 1994)

   -Hypperreactive malarial splenomegaly syndrome (also know as tropical splenomegaly syndrome, which results from poorly understood abnormal immunologic response to malarial infection. May due to loss of suppressor cells and an alteration in the raiton of CD4:CD8 lymphocytes. Typically lack parasitemia and fever on presentation. Requires pancytopenia and IgM elevation for dx. High mortality, and may requity lifelong antimalarial tx))
      -AIDS with MAC infxn,
      -Echinococcal cysts (rarely)

- INFILTRATIVE: Gaucher Disease – lysosomal glucocerebrosidase deficiency. Most common in Ashkenazi Jews, adult onset form exists-typically non-neuropathic in adult form)
   -Niemann-Pick disease- Autosomal recessive lysosomal storage disease. HSM, neuro deficits. Childhood onset

- PORTAL HYPERTENSION

- HEMATOLOGIC- CML, CLL, Hodgkins, heavy chain diseases, amyloid, POEMS (polyneuropathy,organomegaly, endocrinopathy, M protein, and skin lesions-> usually occurs in pts with osteosclerotic multiple myeloma ), Waldenstroms, MM, Myelofibrosis , polycytemia vera