

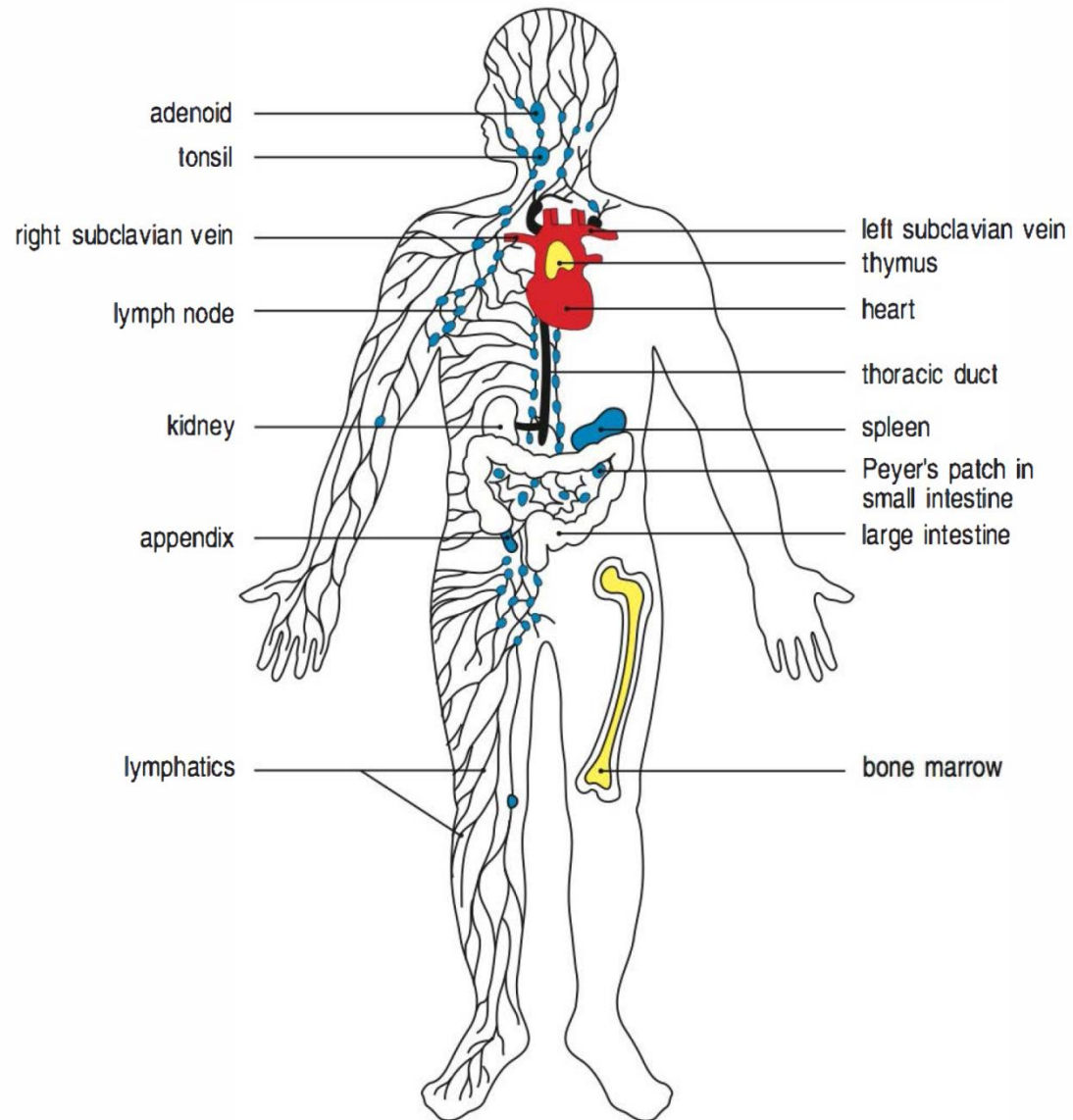
Congenital Asplenia

Case Study

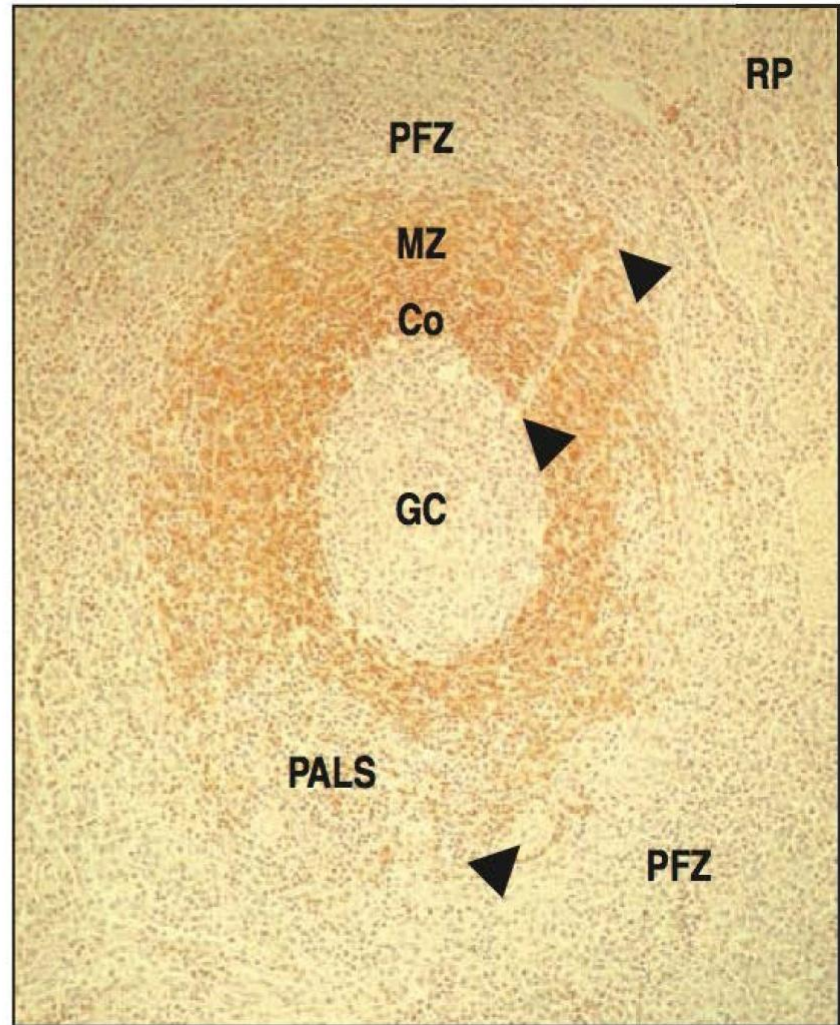
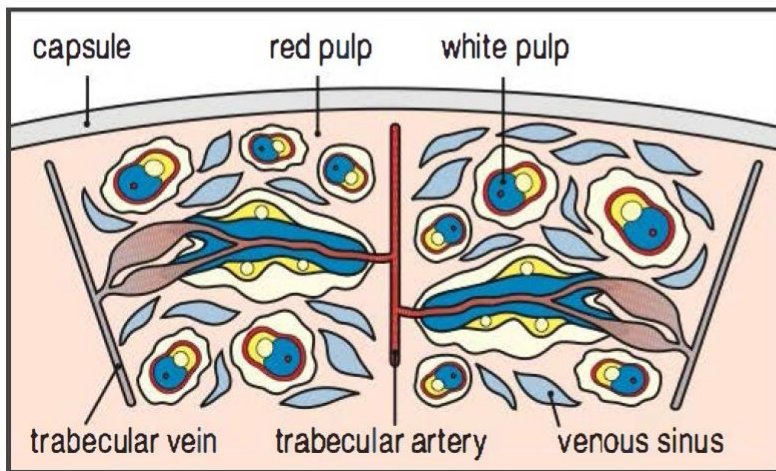
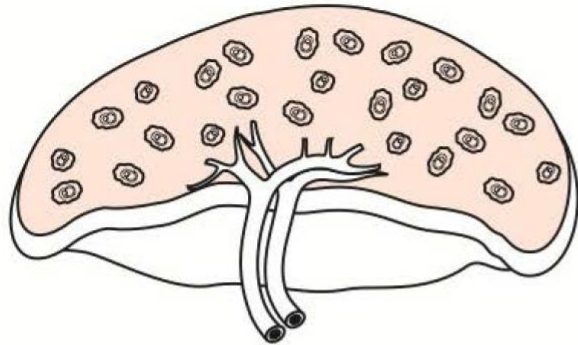
Congenital Asplenia

Unidentified genetic defect.

Baby born with no spleen.



The spleen



Case of Susan Vanderveer

Parents related; consanguineous marriage. 8 kids

5 kids out of 8 had congenital asplenia (Males and females)

High WBC counts, *Haemophilus influenzae* , type b cultures from nose, throat, and blood.

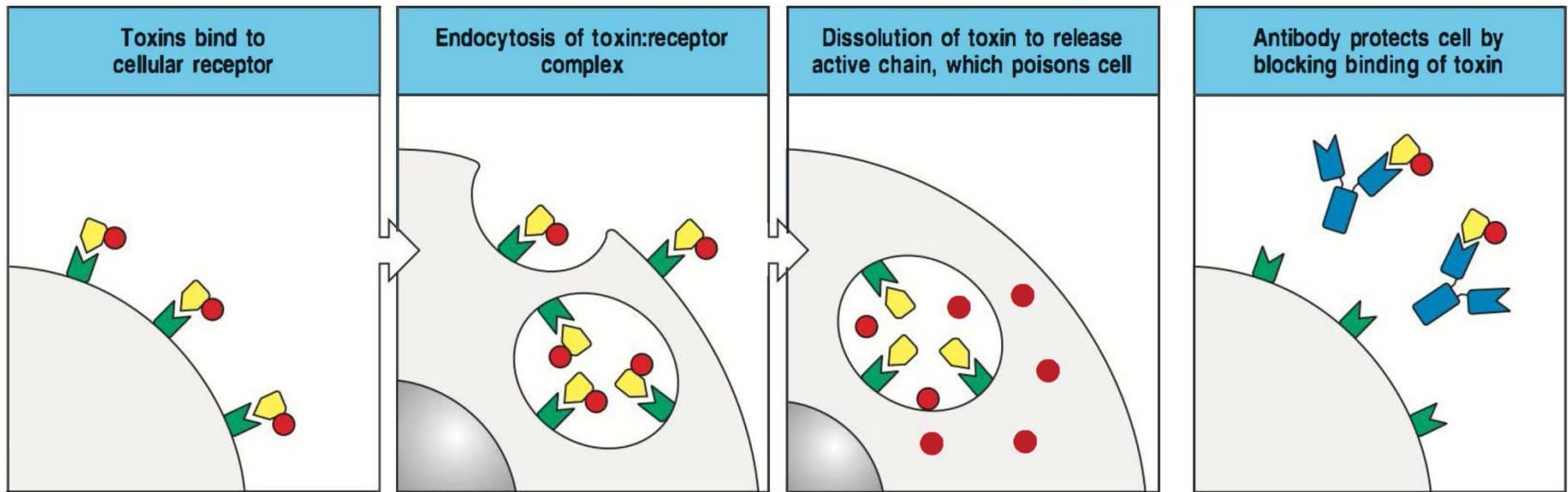
Brother had meningitis caused by *Streptococcus pneumonia*

Kids given DPT and typhoid vaccines and responded to them.

No Ab response to sheep RBCs.

Susan died (10 months) of complications of upper respiratory infection on way to hospital.

Antibodies protect against toxins





A scintillation scan of the abdomen after IV injection
With radioactive colloidal gold Au^{198}

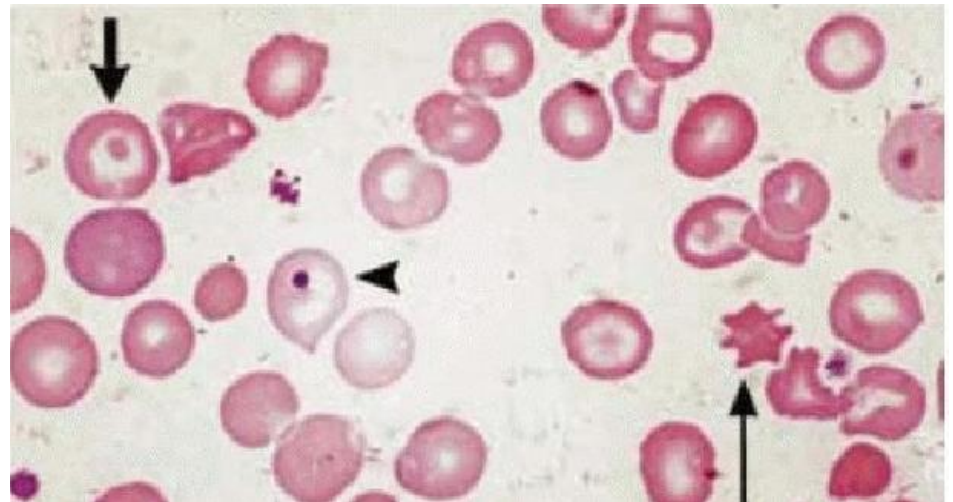


Physical examination

CT, MRI, ultrasonography

Technetium ^{99}Tc Imaging

Peripheral blood film



Why did brother and sister respond to typhoid vaccine but not sheep RBC?

Typhoid vaccine is given s.c. But sheep RBC given IV.

Mode of inheritance of congenital asplenia?

Autosomal recessive

How many kids out of 8 were expected to have the condition?

2

Betsy the sister married a normal man, what is the condition of the kids?

All heterozygous for the defect

5-year-old boy loses spleen in an accident, give consultation?

Check for routine immunizations: DPT, poliovirus, and boosters.

MMR vaccine

Hib Vaccine (*H. Influenzae* type b)

Pneumovax (Vaccine using pneumococcal polysaccharides)

Daily low dose prophylactic antibiotics, higher doses when dental work is done or in case of any invasive surgical procedure.