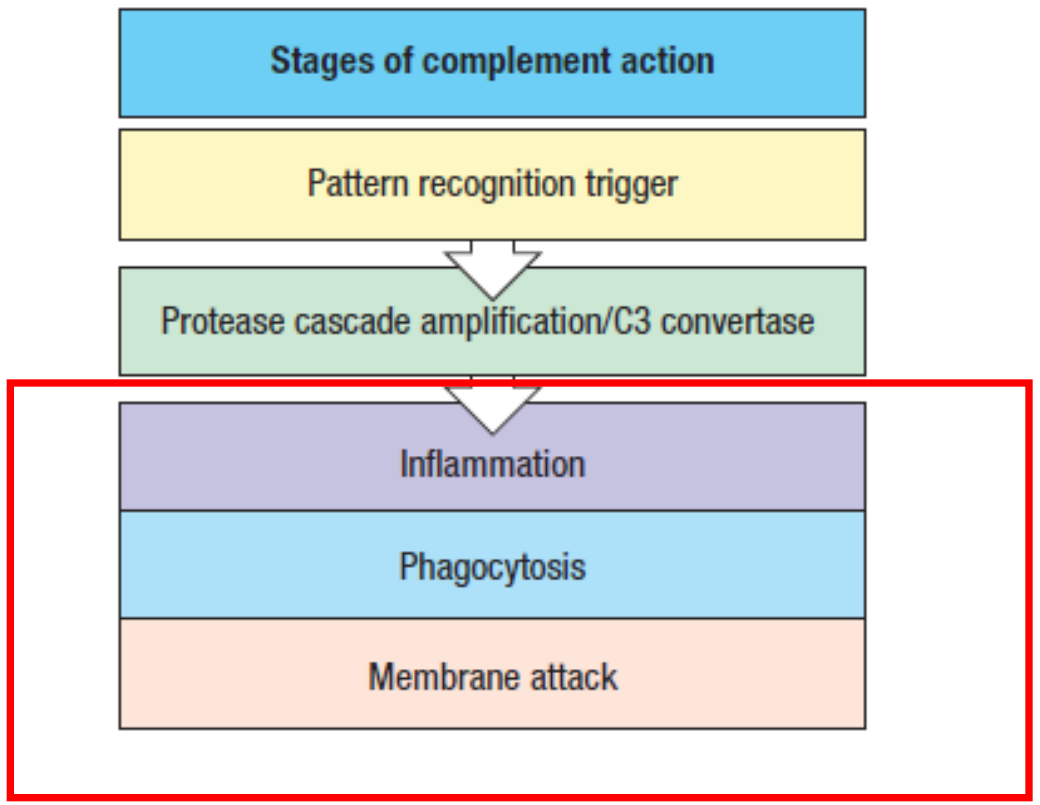
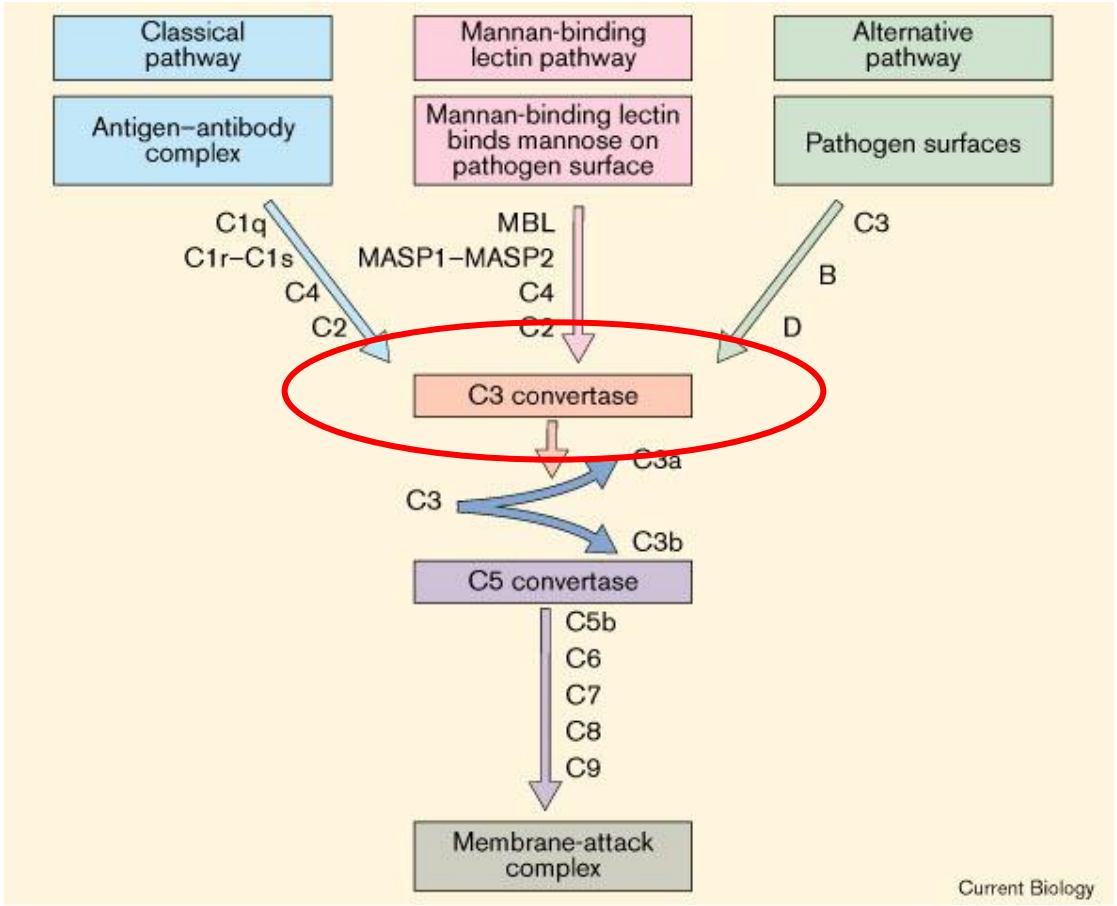
Complement system and immunodeficiencies

What is the complement system and what role does it play in the immune response?

Antibodies serve as detectors of antigens, and the complement system mediates their effector functions.

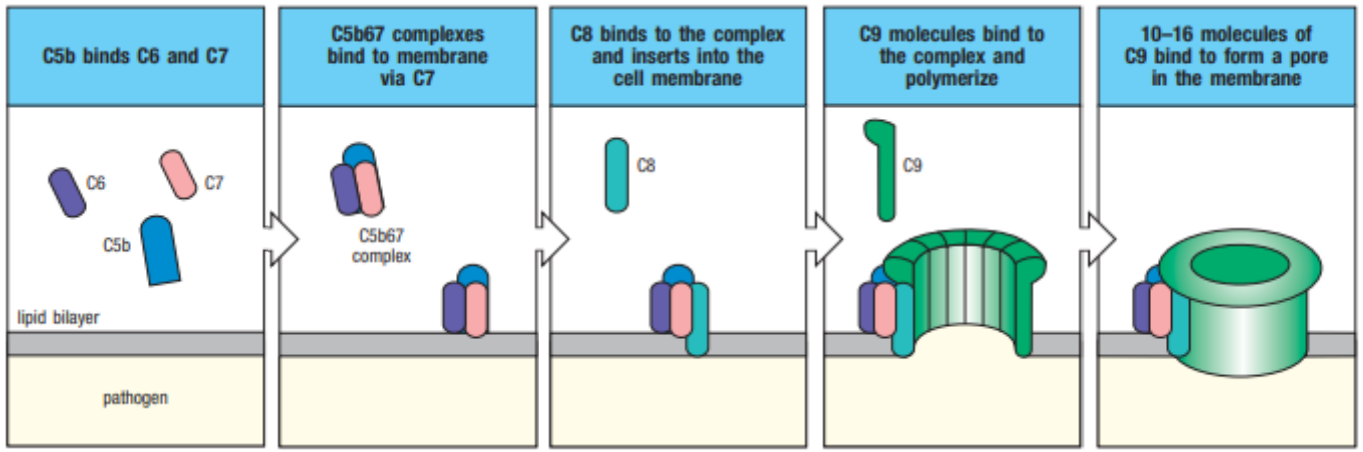


What is the function of C3 convertase in the complement cascade?

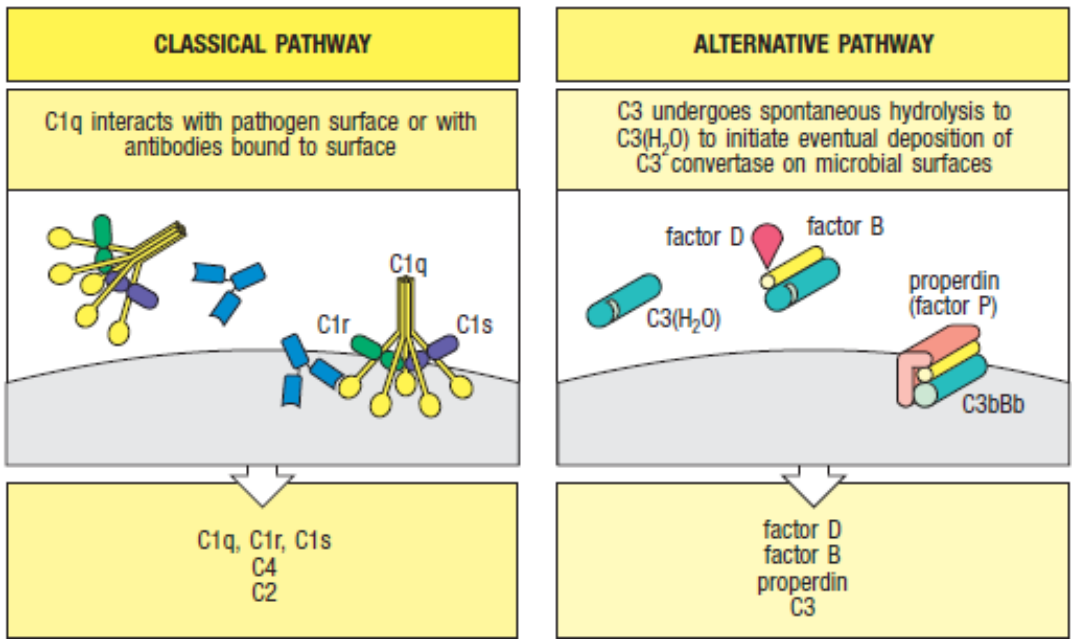


The C3 convertase amplifies the accumulation of complement on the surface of the pathogen and brings the three different initiation mechanisms together to initiate the terminal steps to form the membrane attack complex

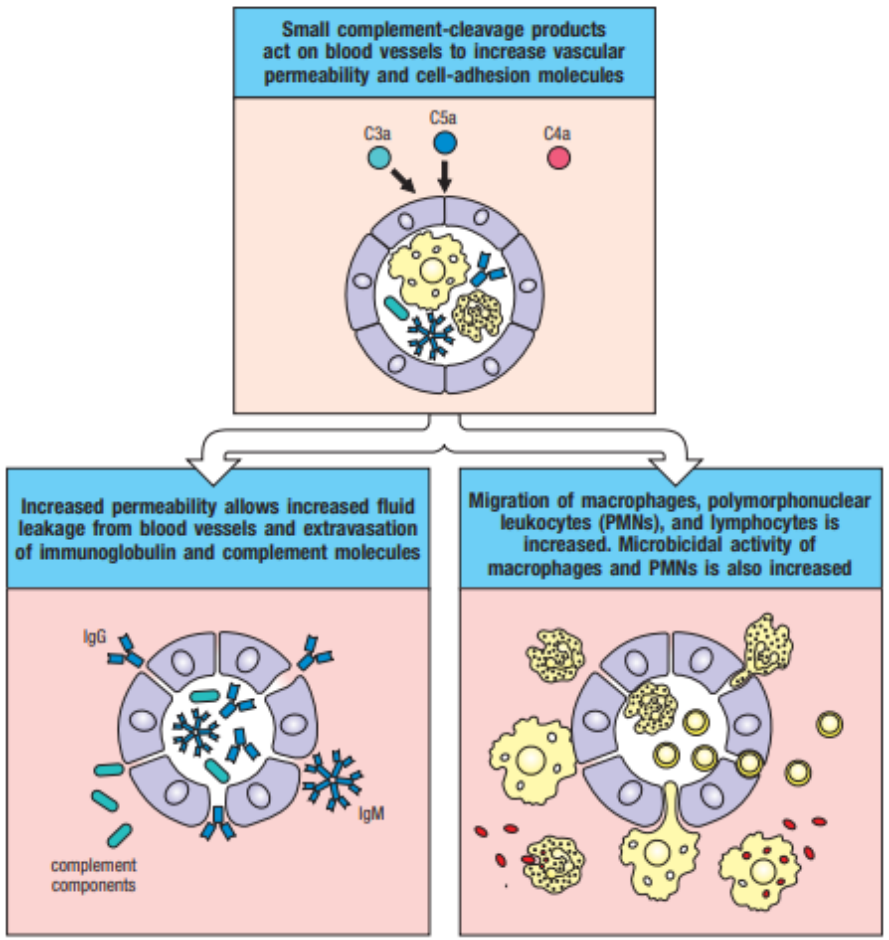
Which complement proteins form the membrane attack complex (MAC)?



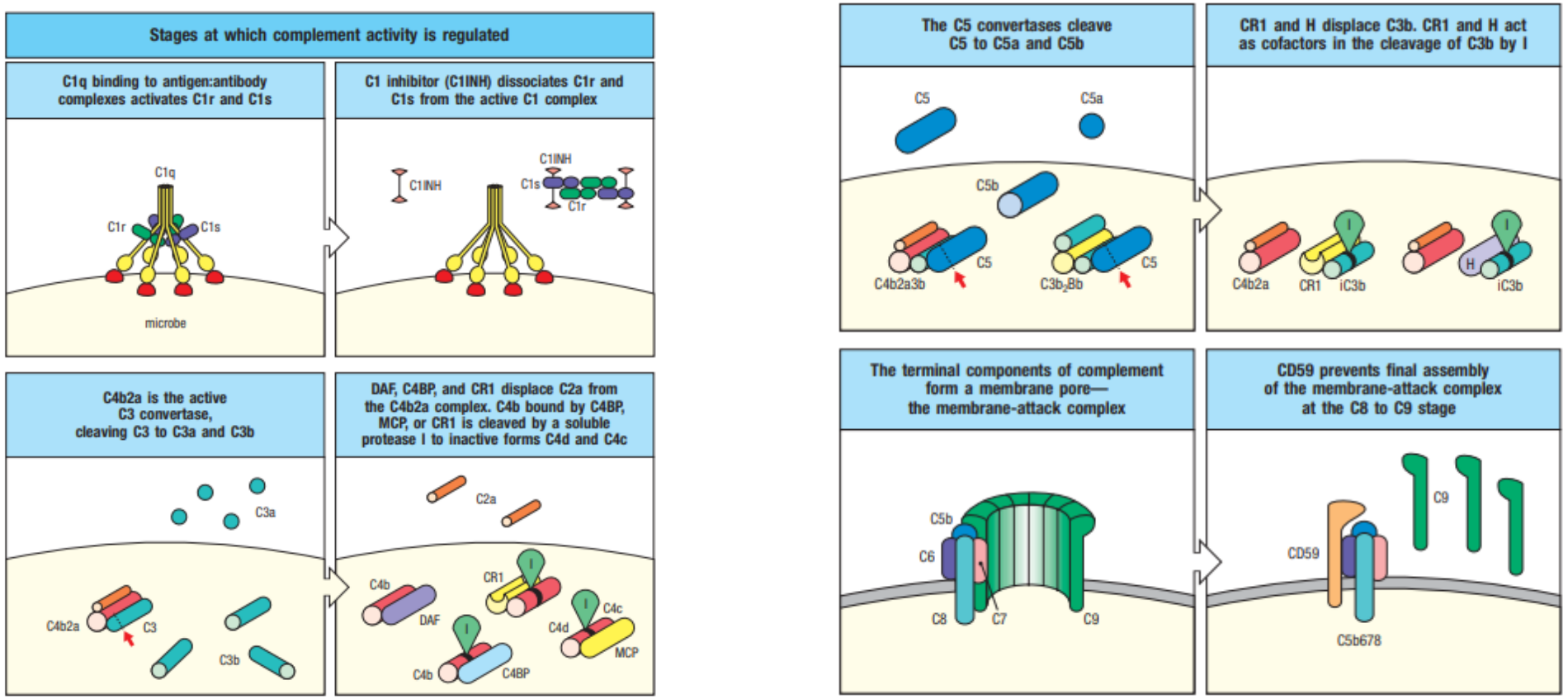
How does the alternative pathway differ from the classical pathway in terms of activation?



What are anaphylatoxins, and which complement fragments are classified as such?



What is the role of complement regulatory proteins in preventing host cell damage?



What is the difference between inherited (primary) and acquired (secondary) immune defects?

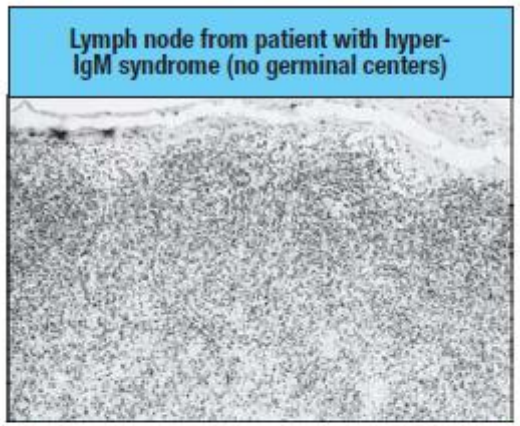
Examples of primary immunodeficiency include:

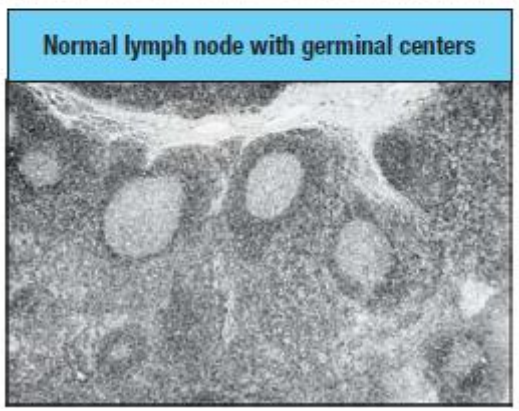
- Severe combined immunodeficiency (SCID).
- Chronic granulomatous disease (CGD).
- Selective IgA deficiency.
- X-linked agammaglobulinemia

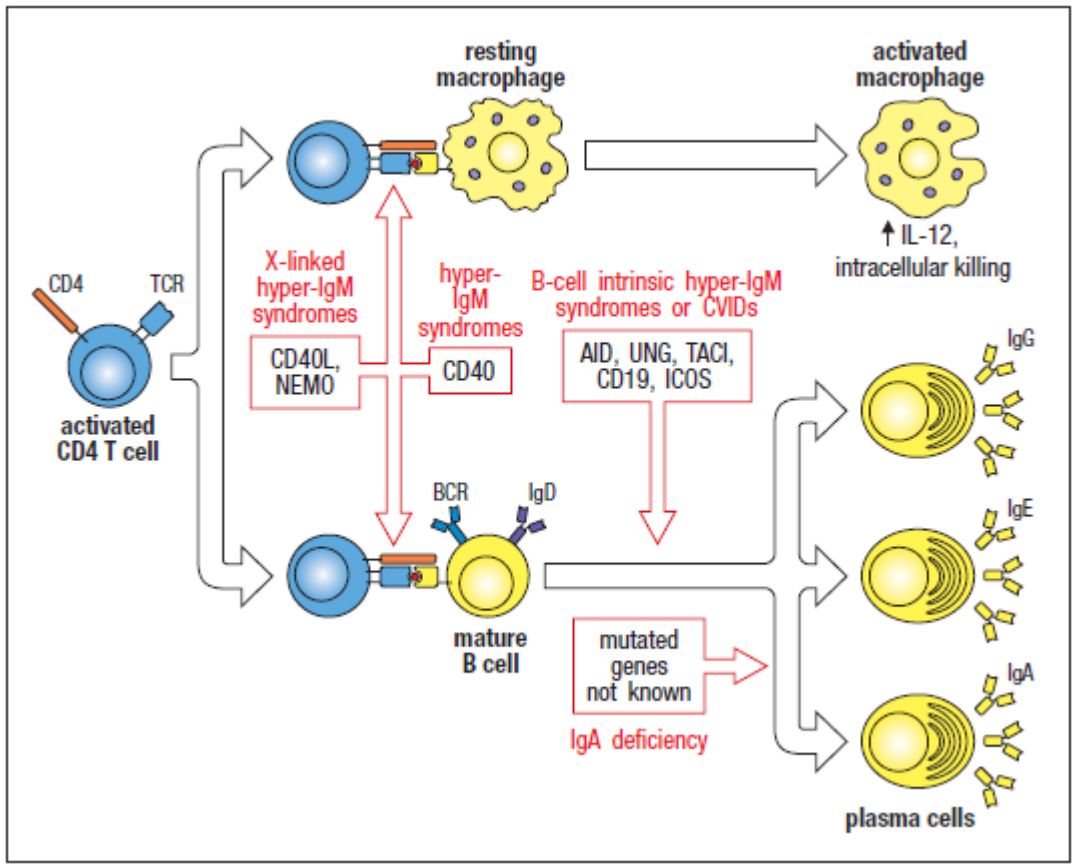
Examples of secondary immunodeficiency disorders include:

- AIDS.
- cancers of the immune system, like leukemia.
- immune-complex diseases, like viral hepatitis.

Name two examples of primary immunodeficiency diseases which affect the function of B-lymphocytes







What genetic mutations are often associated with Severe Combined Immunodeficiency (SCID)?

Name of deficiency syndrome	Specific abnormality	Immune defect	Susceptibility
Severe combined immune deficiency	ADA deficiency	No T or B cells	General
	PNP deficiency	No T or B cells	General
	X-linked *scid*, γ_c chain deficiency	No T cells	General
	Autosomal *scid* DNA repair defect	No T or B cells	General

ADA = adenosine deaminase
PNP = Purine nucleoside phosphorylase
γc chain = common IL-2 receptor gamma chain

How does chemotherapy contribute to acquired immune defects?

Chemotherapy can lead to acquired immune defects primarily by reducing the number of white blood cells, particularly neutrophils

How can bone marrow transplantation help treat certain inherited immune defects?

Bone marrow transplantation (BMT) is a crucial treatment for certain inherited immune defects. By replacing a patient’s defect blood-forming stem cells with healthy ones from a donor, BMT can effectively restore the immune system's ability to fight infections, offering a potential cure for conditions like Severe Combined Immunodeficiency (SCID)

Why are patients with immune defects more susceptible to opportunistic infections?

Immune Defect	Example Disease/Condition	Susceptible Infections/Diseases
T-cell deficiency	SCID	Viruses, fungi, intracellular bacteria
B-cell (antibody) deficiency	X-linked Agammaglobulinemia, CVID	Encapsulated bacteria
Phagocyte defect	CGD, Neutropenia	Catalase-positive bacteria, fungi
Complement deficiency	C3, C5-C9 deficiency	Pyogenic bacteria, Neisseria species
Splenic dysfunction/asplenia	Sickle cell, splenectomy	Encapsulated bacteria

How can HIV infection lead to acquired immune deficiency?