

Severe Combined Immunodeficiency (SCID)

Severe combined immunodeficiency (SCID) is a genetic disorder in which both B and T cells of the immune system are impaired due to one of several possible genes. This disease is sometimes referred to as the bubble boy disease because patients are extremely vulnerable to infectious diseases by viruses, bacteria, fungi, and protozoa and some choose to live in a completely sterile environment. The most common cause of SCID is a defect in the common gamma chain encoded by the gene IL 2 receptor gamma located on the X chromosome. This interleukin receptor plays an important role in the development and differentiation of B and T cells. Furthermore, the defective gamma chain of the receptor is shared by many interleukin receptors, and mutations that result in a non-functional common gamma chain can cause widespread defects in interleukin signaling, leading to almost complete failure of the immune system to develop and function. The second most common form of SCID is caused by a defective adenosine deaminase enzyme, which causes accumulation of dATP which inhibits the activity of ribonucleotide reductase and lymphocyte proliferation. Another cause of SCID is called the Bare lymphocyte syndrome where MHC class II is not expressed on the cell surface of all antigen presenting cells, leading to the failure to synthesize MHC II antigens and inability to mount an appropriate immune response. The most common treatment for SCID is bone marrow transplantation.



PLAY PICMONIC

Both B and T Cell Deficiency

[Basketballs and Tennis-balls](#)

Due to lack of B and T cells, patients are extremely vulnerable to infectious diseases by viruses, bacteria, fungi, and protozoa.

Recurrent Viral, Bacterial, Fungal, Protozoal Infections

[Virus, Bacteria-guy, Fun-guy, Propeller-protozoa Escaping](#)

Patients with Sever Combined Immunodeficiency are susceptible to viral, bacterial, fungal, and protozoal infections.

Defective Interleukin (IL) 2 Receptor

[Broken interlocked \(2\) Tutu receptor](#)

The most common cause of SCID is a defect in the common gamma chain encoded by the gene IL 2 receptor gamma located on the X chromosome. This interleukin receptor plays an important role in the development and differentiation of B and T cells. Furthermore, the defective gamma chain of the receptor is shared by many interleukin receptors, and mutations that result in a non-functional common gamma chain can cause widespread defects in interleukin signaling, leading to almost complete failure of the immune system to develop and function.

X-Linked

[X-suit](#)

The most common cause of SCID is a defect in the common gamma chain encoded by the gene IL 2 receptor gamma located on the X chromosome.

Adenosine Deaminase Deficiency

[A-dentist-sing and Dog-ammo](#)

The second most common form of SCID is caused by a defective adenosine deaminase enzyme, which causes accumulation of dATP which inhibits the activity of ribonucleotide reductase and lymphocyte proliferation.

Failure to Synthesize MHC II Antigens

MHC Complex with (2) Tuts

Another cause of SCID is called the Bare lymphocyte syndrome where MHC class II is not expressed on the cell surface of all antigen presenting cells, leading to the failure to synthesize MHC II antigens and inability to mount an appropriate immune response.

Treat with Bone Marrow Transplant

Bone Train-plant

The most common treatment for SCID is bone marrow transplantation.