Perspective

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Severe Combined Immunodeficiency with Sensitivity to Ionizing Radiation

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Perspective

Combined immunodeficiency also called as combined immune deficiency or CID which is a genetic condition of the immune system. It is known as a primary immunodeficiency. Children inherit the gene for CID from their parents. CID occurs when gene mutations cause defects in the immune system. Severe combined immunodeficiency (SCID) is a group of rare disorders caused by mutations in different genes involved in the development and function of infection-fighting immune cells. Infants with SCID appear healthy at birth but are highly susceptible to severe infections.

The condition is fatal, usually within the first year or two of life, unless infants receive immune-restoring treatments, such as gene therapy, transplants of blood-forming stem cells or enzyme therapy. Development of a newborn screening test has made it possible to detect SCID before symptoms appear, helping ensure that affected infants receive life-saving treatments. Symptoms of SCID occur in infancy and include serious or life-threatening infections which may result in pneumonia and chronic diarrhea. Candida infections of the mouth and diaper area and pneumonia caused by the fungus *Pneumocystis jirovecii* also are common. The SCID newborn screening test originally developed at NIH, measures T cell receptor excision circles (TRECs), a byproduct of T-cell development. Because infants with SCID have few or no T cells, the absence of TRECs may indicate SCID. SCID is caused by genetic a defect that affects the function of T cells.

Depending on the type of SCID, B cells and NK cells can also be affected. These cells play important roles in helping the immune system battle viruses, bacteria, and fungi that cause infections. Affected infants will often die within the first year of life without treatment with hematopoietic stem cell transplantation. Newborn screening for SCID is able to identify infants before they get sick, leading to a shorter time to transplant and offering improved outcomes following transplantation. Transplantation within the first 3 months of life offers the best chances for successful outcomes.

There are several forms of SCID. The most common type is linked to a problem in a gene on the X chromosome, affecting only males. Women may carry the condition, but they also inherit a normal X chromosome. Since starting newborn screening for SCID, recessive forms of the disease that can affect boys and girls have been identified with increased frequency. Severe Combined Immunodeficiency is a serious primary immunodeficiency disease (PI) in which there is combined absence of T lymphocyte and B lymphocyte function. SCID is fatal without a stem cell transplant or corrective gene therapy. There are at least 13 different genetic defects that can cause SCID.

These defects lead to extreme susceptibility to very serious infections. This condition is generally considered to be one of the most serious forms of PI. Effective treatments such as hematopoietic stem cell transplantation, exist that can treat the disorder, and the future holds the promise of gene therapy for some types. SCID is a rare and fatal syndrome of diverse genetic causes in which there is combined absence of T lymphocyte and B lymphocyte function and in many cases also natural killer lymphocyte function. These defects lead to extreme susceptibility to serious infections. There are currently at least thirteen different genes that, when mutated cause SCID.

