Editorial

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Editorial on Warts, Hypogammaglobulinemia, Infections and Myelokathexis (WHIM) Syndrome

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Editorial

Warts, Hypogammaglobulinemia, Infections and Myelokathexis (WHIM) syndrome is a genetic disease characterized by neutropenia, lymphopenia, susceptibility to infections, and myelokathexis, which describes degenerative changes of mature neutrophils and hyperplasia of bone marrow myeloid cells. Some patients present with hypogammaglobulinemia or refractory warts of skin and genitalia. Congenital cardiac defects constitute uncommon manifestations of the disease. The disorder, which is inherited as an autosomal dominant trait, is caused by heterozygous mutations of the chemokine receptor CXCR4. These mutations lead to an increased sensitivity of neutrophils and lymphocytes to the unique ligand CXCL12 and to an increased accumulation of mature neutrophils in the bone marrow.

Warts, Hypogammaglobulinemia, Infections, and Myelokathexis (WHIM) syndrome is a rare genetic disease of the immune system. Its name is an acronym for its main clinical manifestations: warts, hypogammaglobulinemia, infections, and myelokathexis. Hypogammaglobulinemia is a deficiency in specific infection-fighting antibodies in the blood. Myelokathexis refers to the failure of neutrophils — infection-fighting white blood cells — to move from the bone marrow into the bloodstream where they can patrol the body. WHIM syndrome patients also have trouble distributing most other types of immune cells to the blood. Such defects in the immune system predispose WHIM syndrome patients to frequent bacterial and viral infections, persistent skin and genital warts, and an increased risk of developing cancer caused by human papillomavirus.

Standard therapy for WHIM syndrome aims to restore deficient blood components. It involves intravenous immunoglobulin, a blood product containing antibodies, or Granulocyte Colony Stimulating Factor (G-CSF), an immune-cell-growth molecule. However, these treatments do not specifically target the CXCR4 genetic defect, and their long-term efficacy has not been established through clinical trials. WHIM syndrome is a rare, autosomal dominant, combined immunodeficiency with a complex and variable phenotype. WHIM is an acronym for the four major clinical manifestations of the syndrome: warts, hypogammaglobulinemia, infections, and myelokathexis. Myelokathexis is the bone marrow sequestration of mature neutrophils which occurs because of mutations that increase the function of the chemokine receptor CXCR4. However, most other leukocytes also express CXCR4, and there is panleukopenia with low numbers of circulating monocytes, T cells, and B cells.

The symptoms of WHIM syndrome can vary greatly from one person to another. Some may only have mild symptoms; others may develop potentially life-threatening complications. Because of the small number of identified cases, the lack of large clinical studies, and the possibility of other genes influencing the disease outcome, health care providers cannot develop an accurate picture of associated symptoms and prognosis. Generally, symptoms first appear in early childhood when most children with WHIM syndrome experience repeated bacterial infections that can be mild or severe, but usually respond promptly to antibiotic therapy. The number and frequency of infections can vary greatly from one child to another.

