WHIM Syndrome

A rare immunodeficiency whose name spells its four clues (CXCR4 mutation)

W — **Warts**
extensive, resistant HPV warts

H — **Hypogammaglobulinemia**
low antibody (humoral) defenses

I — **Infections**
recurrent bacterial infections

M — **Myelokathexis**
neutrophils "trapped" in the bone marrow, with neutropenia in blood

Cause	The paradox	Treatment
Gain-of-function CXCR4 mutation	Neutropenia with a full bone marrow	Antibiotics, IVIG, G-CSF (plerixafor)

Treatment: antibiotics, immunoglobulin replacement and colony-stimulating factors (G-CSF) to mobilize neutrophils