# Combined immunodeficiency due to CD27 deficiency

INSERM

## Source

*INSERM. (1999). Orphanet: an online rare disease and orphan drug data base. Combined immunodeficiency due to CD27 deficiency. ORPHA:238505*

Autosomal recessive lymphoproliferative disease is a rare combined T and B cell immunodeficiency with a predisposition to lymphoproliferative syndrome. It is characterized by persistent symptomatic EBV-viremia and hypogammaglobulinemia variably presenting with fever, lymphadenopathy and systemic inflammatory conditions including hepatitis, pneumonia and sepsis. It may be associated with lymphoma, hemophagocytic lymphohistiocytosis, and aplastic anemia.